Third Edition

CRANIAL NERVES

FUNCTION & DYSFUNCTION

L. Wilson-Pauwels • P. A. Stewart
E. J. Akesson • S. D. Spacey

CRANIAL NERVES

Function and Dysfunction

THIRD EDITION

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To the memory of Tommy;
may his spirit soar forever

Preface

The third edition of *Cranial Nerves*, titled *Cranial Nerves: Function & Dysfunction*, continues to present problem-based learning cases and clinical testing in a visual format. This book has maintained the original approach of the first edition; the text and figures integrate important concepts related to the neuroanatomy and gross anatomy of the cranial nerves. The user is guided through pertinent information and color-coded functional drawings of the pathways/modalities from the periphery of the body to the brain (sensory input) and from the brain to the periphery (motor output). Each pathway is described according to the direction of the nerve impulse and not according to the embryologic outgrowth of the nerve. Through the separation of nerve fiber modalities, we highlight the important clinical aspects of each nerve.

In this edition, we have significantly updated the chapters in keeping with recent advances in basic science and in clinical medicine. Many of the figures have been enhanced, and many are new, with the goal of visually communicating the route of each cranial nerve. The link to the *Cranial Nerve* website includes new, high-quality clinical testing videos (http://www.pmph-usa.com/cranialnerves).

Cranial Nerves is still targeted to students of the health sciences (medicine, rehabilitation sciences, dentistry, pharmacy, speech pathology, audiology, nursing, physical and health education, and biomedical communications) who may be studying neuroanatomy and gross anatomy for the first time. Since the first edition (printed in 1988), we have discovered that the book is also a valuable quick reference for residents in neurology, neurosurgery, otolaryngology, and maxillofacial surgery.

We are pleased that this third edition of *Cranial Nerves* is published by PMPH (Peoples Medical Publishing House) USA and we would like to take this opportunity to thank Mr. Jason Malley, Executive Editor, for his support in the development of this new edition. We also extend a special thanks to his colleagues at PMPH USA for their creative designs and their expertise in the editing of this edition.

The following list includes the names of our colleagues who have offered their time and expertise to review aspects of the text and figures of the three editions of *Cranial Nerves*. We are very appreciative of their contributions to this publication.

Linda Wilson-Pauwels
Patricia Stewart
Elizabeth Akesson
Siân Spacey

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Our special thanks to our following colleagues from Biomedical Communications, University of Toronto; Professor Jodie Jenkinson, the videographer and instruc-

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We are especially grateful to our student readers. They are: from the University of Toronto, Ms. Melissa Bayne and Mr. Simon McVaugh-Smock, Department of Speech Language Pathology, and Ms. Allison Guy, Faculty of Arts and Science; and from the University of British Columbia, Ms. Maria Glavas from the Department of Anatomy.

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We are especially indebted to Dr. Wayne Vogl who read and critiqued many chapters of this book. His input was invaluable.

We are appreciative of the excellent videos and photographs produced by Mr. Mark Fenger as well as our model Dr. Ryan Punambolam in the videos and photographs.

We also thank the following physicians who provided medical images to supplement our case histories: Dr. Robert Nugent, Vancouver General Hospital, University of British Columbia (Figure I–1); Dr. Ray Buncic (Figures II–4 and II–19); Cynthia VandenHoven, The Hospital for Sick Children, Toronto (Figure II–4); Dr. David Mikulis, Brain Imaging & Behaviour Systems—Neuroscience, Toronto Western Research Institute (Figure V–16); and Dr. D. S. Butcher, William Osler Health Centre, Brampton, Ontario (Figure VIII–13).

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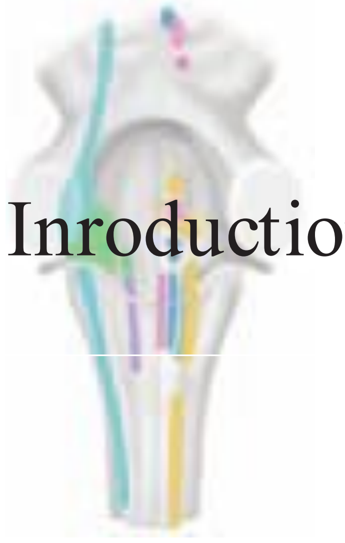
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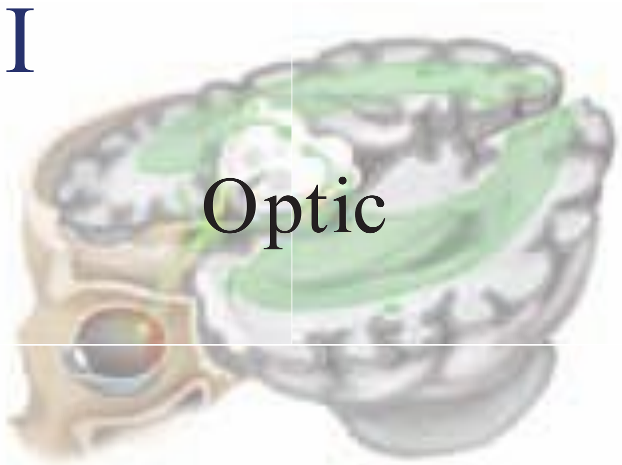
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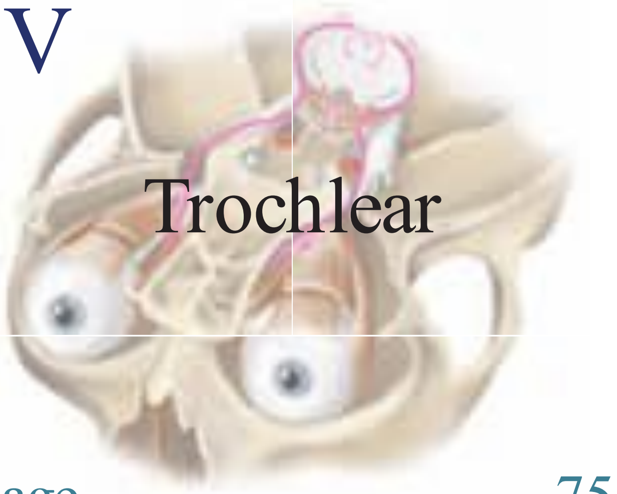
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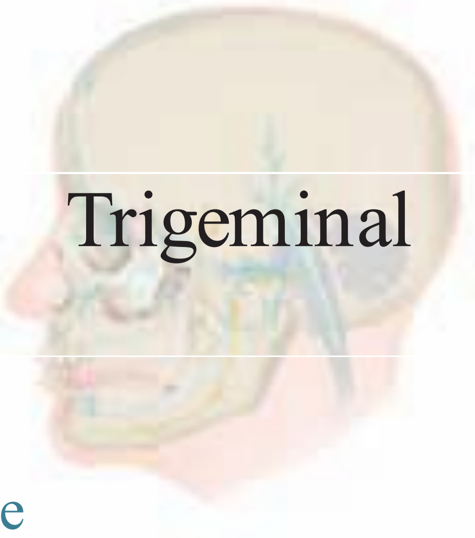
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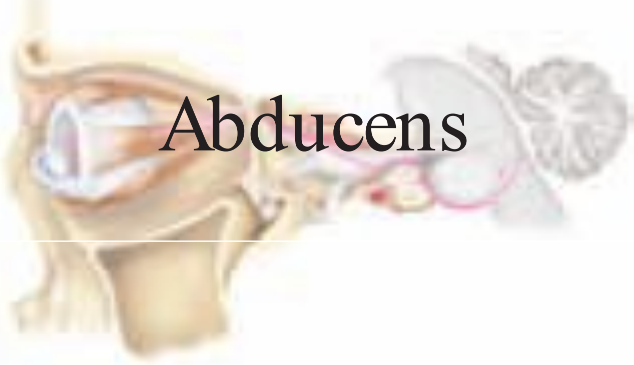
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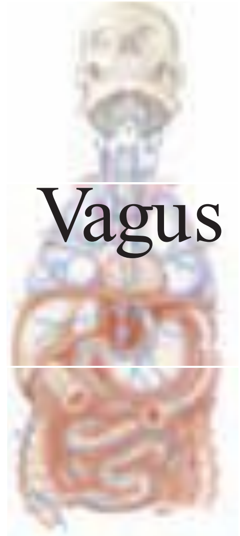
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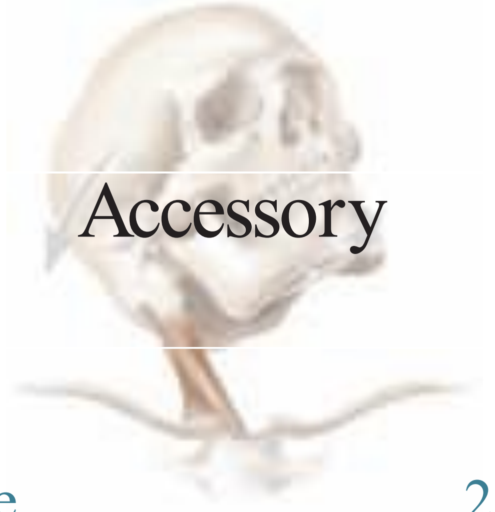
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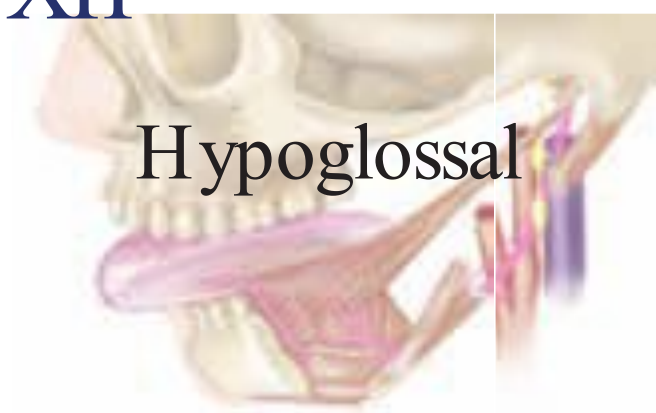
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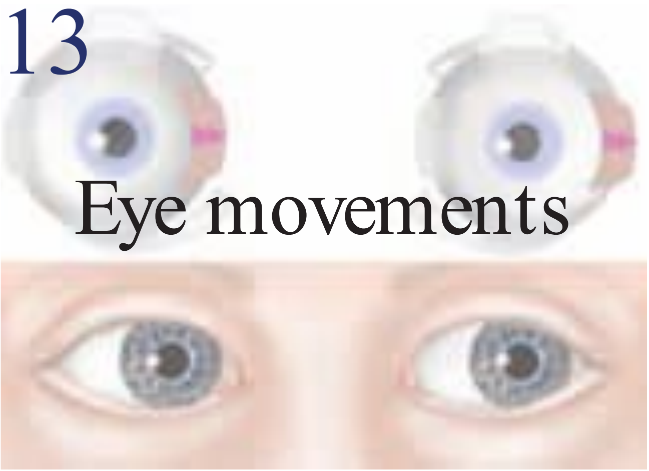
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Introduction

The twelve pairs of cranial nerves provide sensory and motor innervation for the head and neck including general, visceral, and special sensory; and voluntary somatic, branchial, and involuntary parasympathetic motor control. Because these nerves emerge from the cranium, they are called *cranial* nerves, as opposed to spinal nerves that emerge from the spinal column.

The cranial nerves function as modified spinal nerves. As a group, they have both sensory and motor* components; however, individual nerves may be purely sensory, purely motor, or mixed (both motor and sensory).

The cranial nerves carry *six distinct modalities*—three sensory and three motor (Figure Intro–1 and Tables Intro–1 and Intro–2). These modalities are

- **General sensory nerves,** which mediate touch, pain, temperature, pressure, vibration, and proprioceptive sensation.
- **Visceral sensory nerves,** which mediate sensory input, except pain, from the viscera.
- **Special sensory nerves,** which mediate smell, vision, taste, hearing, and balance.
- **Somatic motor nerves,** which innervate the voluntary muscles that develop from the somites.
- **Branchial motor nerves,** which innervate the voluntary muscles that develop from the branchial arches.
- **Parasympathetic (visceral) motor nerves,** which innervate the viscera, including glands, bodies, mucosae, and all involuntary smooth muscle.

---

*In this text, we have chosen to use the terms “sensory” and “motor” rather than the terms “efferent” and “afferent,” which are internationally recognized and detailed in *Terminologia Anatomica*. In written work, the use of “afferent” and “efferent” appeals to scholars because it avoids the difficulties in defining “motor” and “sensory” by describing only the direction of the impulse. In lectures, however, “afferent” and “efferent” sound so much alike that students find them difficult to distinguish, and we have found their use to be confusing.

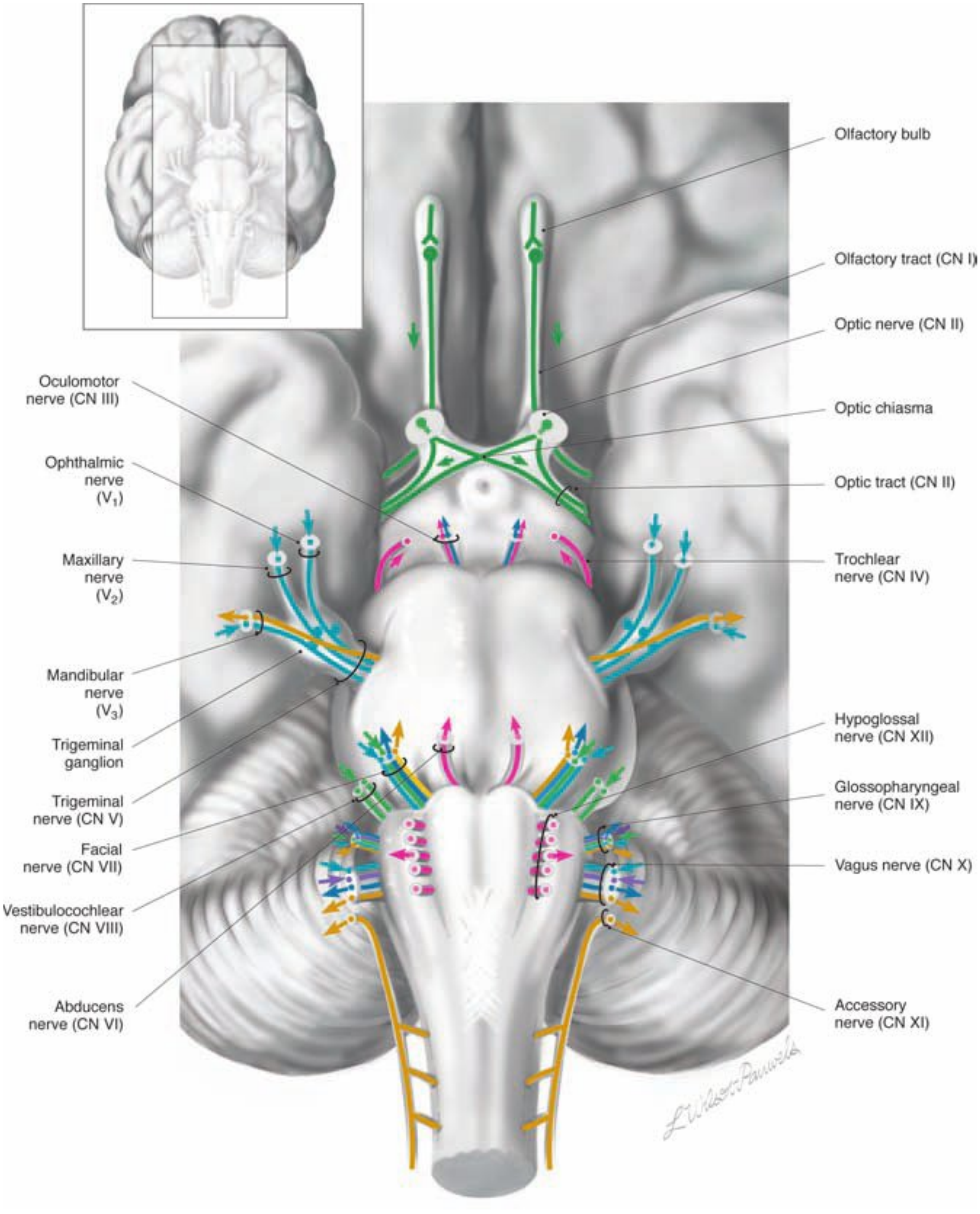


FIGURE Intro–1 Basal view of the brain and brain stem (cropped).

TABLE Intro–1 Cranial Nerves: Components and Functions

	General	Visceral	Special	Somatic	Branchial	Para.	(For more detail see appropriate chapter)
Olfactory CN I			☐				Special sensory for smell
Optic CN II			☐				Special sensory for vision
Oculomotor CN III				☐			Somatic motor to all extraocular muscles except superior oblique and lateral rectus muscles
						☐	Parasympathetic motor to ciliary and constrictor pupillae muscles
Trochlear CN IV				☐			Somatic motor to superior oblique muscle
Trigeminal CN V	☐						General sensory from face, anterior scalp as far posterior as the apex of the head, conjunctiva, bulb of the eye, mucous membranes of paranasal sinuses, nasal and oral cavities including the tongue and teeth, part of the external aspect of the tympanic membrane, and meninges of anterior and middle cranial fossae
					☐		Branchial motor to muscles of mastication (masseter, temporalis, medial and lateral pterygoid), tensores tympani, tensores veli palatini, mylohyoid, and anterior belly of the digastric muscles
Abducens CN VI				☐			Somatic motor to lateral rectus muscle
Facial CN VII	☐						General sensory from a small variable area of the concha of the external ear (pinna/auricle), external acoustic meatus, external (lateral) surface of the tympanic membrane, and a small area of skin behind the ear
			☐				Special sensory for taste from anterior two-thirds of tongue and soft palate
					☐		Branchial motor to muscles of facial expression
						☐	Parasympathetic motor to lacrimal, submandibular. and sublingual glands, and oral, nasal, and pharyngeal mucosal glands

(continued)

TABLE Intro–1 Cranial Nerves: Components and Functions (continued)

	General	Visceral	Special	Somatic	Branchial	Para.	(For more detail see appropriate chapter)
Vestibulocochlear CN VIII							
Vestibular division			☐				Special sensory for balance
Cochlear division			☐				Special sensory for hearing
Glossopharyngeal CN IX	☐						General sensory from posterior one-third of tongue, tonsil, soft palate, fauces, uvula, mucosa of the internal surface of tympanic membrane and cavity, mastoid air cells, auditory tube, and upper pharynx
		☐					Visceral sensory from the carotid body and sinus
			☐				Special sensory for taste from posterior one-third of tongue
					☐		Branchial motor to stylopharyngeus muscle
						☐	Parasympathetic motor to parotid gland and blood vessels in carotid body and sinus
Vagus CN X	☐						General sensory from posterior meninges, concha, skin at the back of the ear and in external acoustic meatus, part of the external surface of the tympanic membrane and pharynx, and larynx
		☐					Visceral sensory from lower pharynx, larynx, trachea (caudal part), esophagus, and thoracic and abdominal viscera, stretch receptors in walls of the aortic arch and chemoreceptors in aortic bodies
					☐		Branchial motor to superior, middle, and inferior constrictors, levator veli palati, salpingopharyngeus, palatopharyngeus, palatoglossus, and intrinsic muscles of the larynx
						☐	Parasympathetic motor to smooth muscles and glands of the pharynx, larynx, thoracic and abdominal viscera and cardiac muscle
Accessory CN XI					☐		Branchial motor to sternomastoid and upper fibers of trapezius muscles
HypoglossalCN XII				☐			Somatic motor to intrinsic muscles of the tongue and extrinsic muscles of the tongue except palatoglossus

TABLE Intro–2 Components, Cranial Nerves, Nuclei, and Functions

Component	Nerve	Nucleus/Cells	Function
General sensory (afferent)	CN V	Trigeminal	Face, anterior scalp as far posterior as the apex of the head, conjunctiva, bulb of the eye, mucous membranes of paranasal sinuses, nasal and oral cavities including the tongue and teeth, part of the external aspect of the tympanic membrane, and meninges of anterior and middle cranial fossae
	CN VII	Trigeminal	Small variable area of the concha of the external ear (pinna/auricle), external acoustic meatus, external (lateral) surface of the tympanic membrane, and a small area of skin behind the ear
	CN IX	Trigeminal	Posterior one-third of tongue, tonsil, soft palate, fauces, uvula, mucosa of the internal surface of tympanic membrane and cavity, mastoid air cells, auditory tube, and upper pharynx
	CN X	Trigeminal	Posterior meninges, concha, skin at the back of the ear and in external acoustic meatus, part of the external surface of the tympanic membrane and pharynx, and larynx
Visceral sensory (afferent)	CN IX	Solitarius*	Carotid body and sinus
	CN X	Solitarius*	Lower pharynx, larynx, trachea (caudal part), esophagus, and thoracic and abdominal viscera, stretch receptors in walls of the aortic arch and chemoreceptors in the aortic bodies
Special sensory (afferent)†	CN I	Olfactory bulb	Smell
	CN II	Ganglion cells of the retina	Vision
	CN VII	Gustatory nucleus‡	Taste—anterior two-thirds of tongue and soft palete
	CN VIII	Vestibular nucleus	Balance
	CN VIII	Cochlear nucleus	Hearing

(continued)

TABLE Intro–2 Components, Cranial Nerves, Nuclei, and Functions (continued)

Component	Nerve	Nucleus/Cells	Function
	CN IX	Gustatory nucleus‡	Taste—posterior one-third of tongue
Somatic motor (efferent)	CN III	Oculomotor	All extraocular eye muscles except superior oblique and lateral rectus
	CN IV	Trochlear	Superior oblique muscle
	CN VI	Abducens	Lateral rectus muscle
	CN XII	Hypoglossal	Intrinsic and extrinsic tongue muscles except palatoglossus
Branchial motor (efferent)	CN V	Masticator	Muscles of mastication (masseter, temporalis, medial and lateral pterygoid), tensores tympani, tensores veli palatini, mylohyoid, and anterior belly of the digastric muscles
	CN VII	Facial motor	Muscles of facial expression
	CN IX	Ambiguus	Stylopharyngeus muscle
	CN X	Ambiguus	Superior, middle, and inferior constrictors, levator palati, salpingopharyngeus, palatopharyngeus, palatoglossus, and intrinsic muscles of the larynx
	CN XI	Accessory§	Sternomastoid and upper trapezius muscles
Parasympathetic (visceral) motor	CN III	Edinger-Westphal	Ciliary and constrictor pupillae muscles
	CN VII	Superior salivatory	Lacrimal, submandibular and sublingual glands, and oral, nasal and pharyngeal mucosal glands
	CN IX	Inferior salivatory	Parotid gland
		Ambiguus	Blood vessels in carotid body and sinus
	CN X	Dorsal vagal	Smooth muscles and glands of the pharynx, larynx, and thoracic and abdominal viscera
	CN X	Ambiguus	Cardiac muscle

*More properly known as the "nucleus of the tractus solitarius."

†Special sensory nuclei are defined as the cell bodies of the secondary sensory neurons.

‡The gustatory nucleus is the rostral portion of the nucleus of the tractus solitarius.

§In this text, we do not follow the convention of identifying the caudal fibers of CN X that run briefly with CN XI as the "cranial root of XI" (see Chapter X for further discussion).

CN, cranial nerve.

In this book, each nerve modality has been assigned a different color; the color scheme is adhered to throughout. Figure Intro–1 provides an overview of the base of the brain and brain stem with the cranial nerves in situ. Tables Intro–1 and Intro–2 provide a summary of the cranial nerves, their modalities, and their functions. A bookmark on the back cover flap (Fig. Intro–2) provides an easy reference for the color coding of the tracts. As you are reading the book, place the flap over the illustrations to identify each modality.

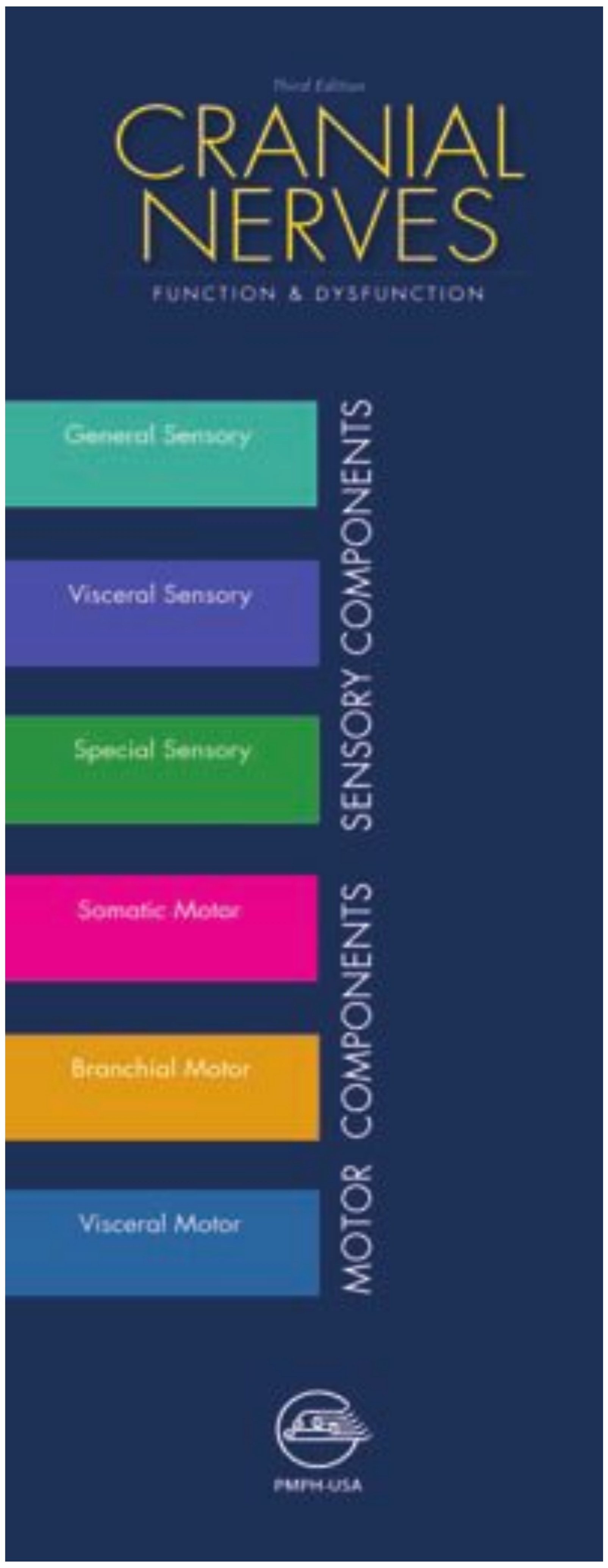


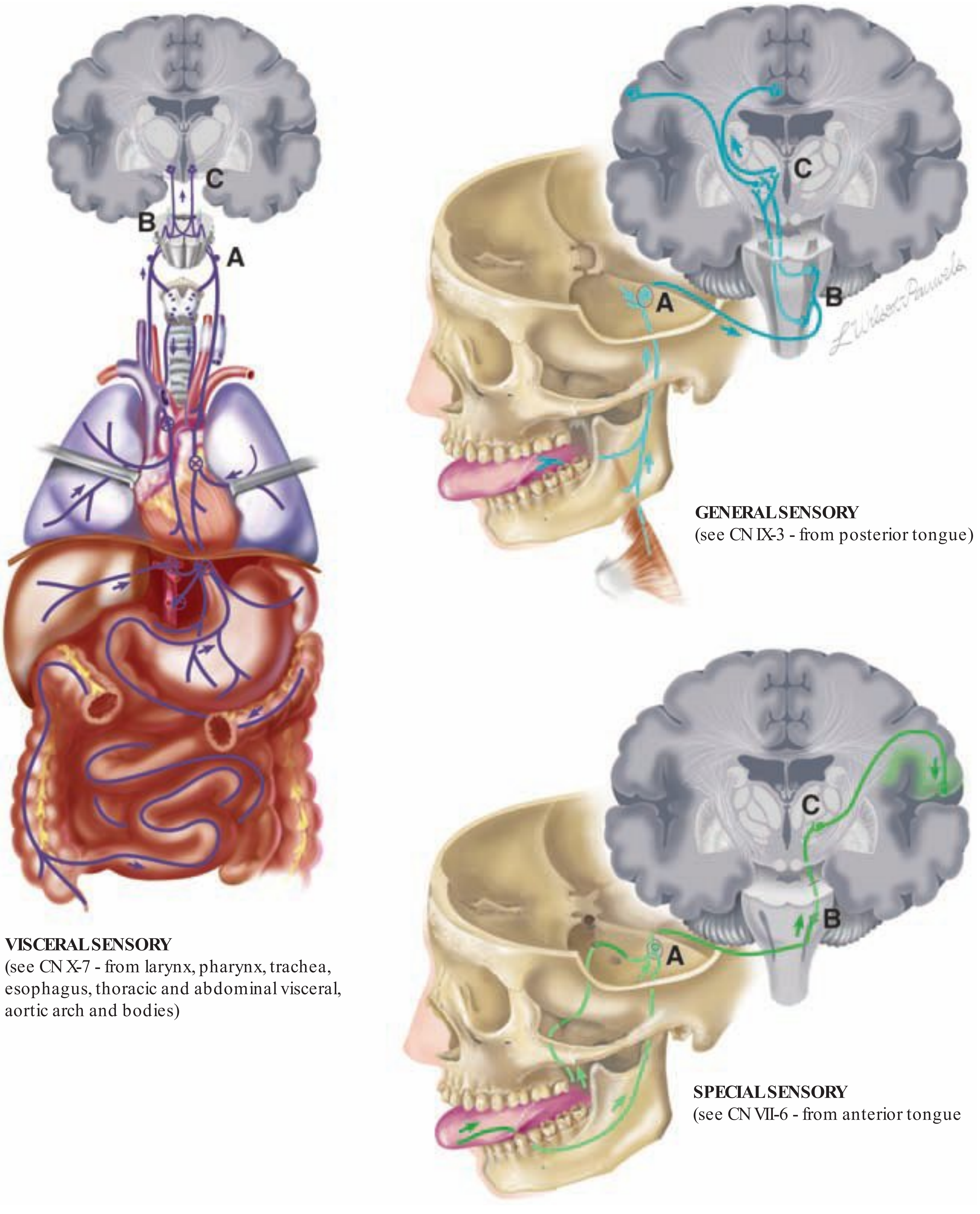
FIGURE Intro–2 Bookmark on back cover

GENERAL, VISCERAL, AND SPECIAL SENSORY (AFFERENT) PATHWAYS OF CRANIAL NERVES

Sensory pathways are composed of three major neurons: primary, secondary, and tertiary. The locations of the major neurons in the pathways for different sensory modalities are illustrated in Figure Intro–3.

1. The **primary sensory neuron** cell bodies are usually located outside the central nervous system (CNS) in sensory ganglia. They are homologous with the dorsal root ganglia of the spinal cord but are usually smaller and are frequently overlooked.
2. The **secondary sensory neuron** cell bodies are located in the dorsal and lateral gray matter of the brain stem, and their axons usually cross the midline to project to the thalamus. The cell bodies that reside in the brain stem form the sensory group of cranial nerve nuclei.
3. The **tertiary sensory neuron** cell bodies are located in the thalamus, and their axons project to the sensory cortex.

The sensory component of the cranial nerves, except for cranial nerves (CNs) I and II, consists of the axons of the primary sensory neurons. CNs I and II are special cases that will be explained in the appropriate chapters. Because there are several modalities carried by sensory neurons, and because these modalities tend to follow



VISCERAL SENSORY
(see CN X-7 - from larynx, pharynx, trachea, esophagus, thoracic and abdominal visceral, aortic arch and bodies)

GENERAL SENSORY
(see CN IX-3 - from posterior tongue)

SPECIAL SENSORY
(see CN VII-6 - from anterior tongue

FIGURE Intro–3 Examples of sensory (afferent) pathways: **General**, **Visceral** and **Special**
A. Primary neuron cell body
B. Secondary neuron cell body
C. Tertiary neuron cell body

different pathways in the brain stem, the loss experienced when sensory neurons are damaged depends, to a large extent, on the location of the lesion.

- Lesions in a peripheral nerve result in the loss of all sensation carried by that nerve from its field of distribution.
- Sensory abnormalities resulting from lesions in the CNS depend on which sensory pathways are affected. For example, a lesion in the descending portion of the trigeminal nucleus results in loss of pain and temperature sensation on the affected side of the face but in little loss of discriminative touch, which is perceived in the middle and upper part of the same nucleus (see Chapter V, Figs. V–12 and V–13).
- Damage to the thalamus results in a patchy hemianesthesia (numbness) and hemianalgesia (insensitivity to pain) on the contralateral (opposite) side of the body. There is often additional spontaneous pain of an unpleasant, disturbing nature on the partially anesthetized side.

SOMATIC, BRANCHIAL AND PARASYMPATHETIC MOTOR (EFFERENT) PATHWAYS OF CRANIAL NERVES

Voluntary motor pathways (somatic and branchial) are composed of two major neurons: the upper motor neuron and the lower motor neuron. The involuntary motor pathway (parasympathetic) is composed of three major neurons: the upper motor neuron, the lower motor neuron, and the tertiary motor neuron within a ganglion or body.

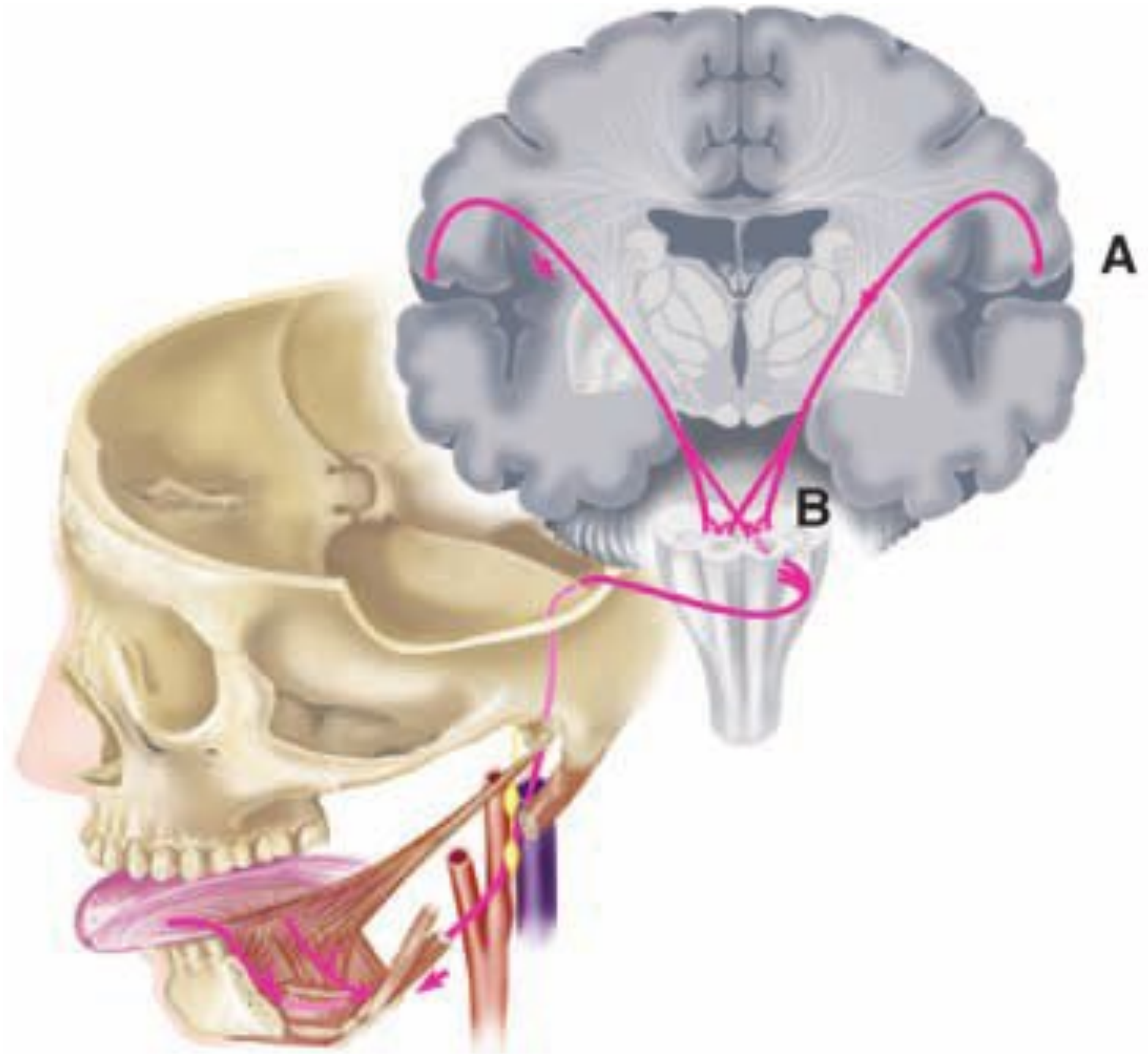
Examples of the somatic and branchial motor pathways are illustrated in Figure Intro–4.

1. The **upper motor neuron** is usually located in the cerebral cortex. Its axon projects caudally through the corticobulbar tract[†] to contact the lower motor neuron in the brain stem. Most, but not all, of the motor pathways that terminate in the brain stem project bilaterally to contact lower motor neurons on both sides of the midline.

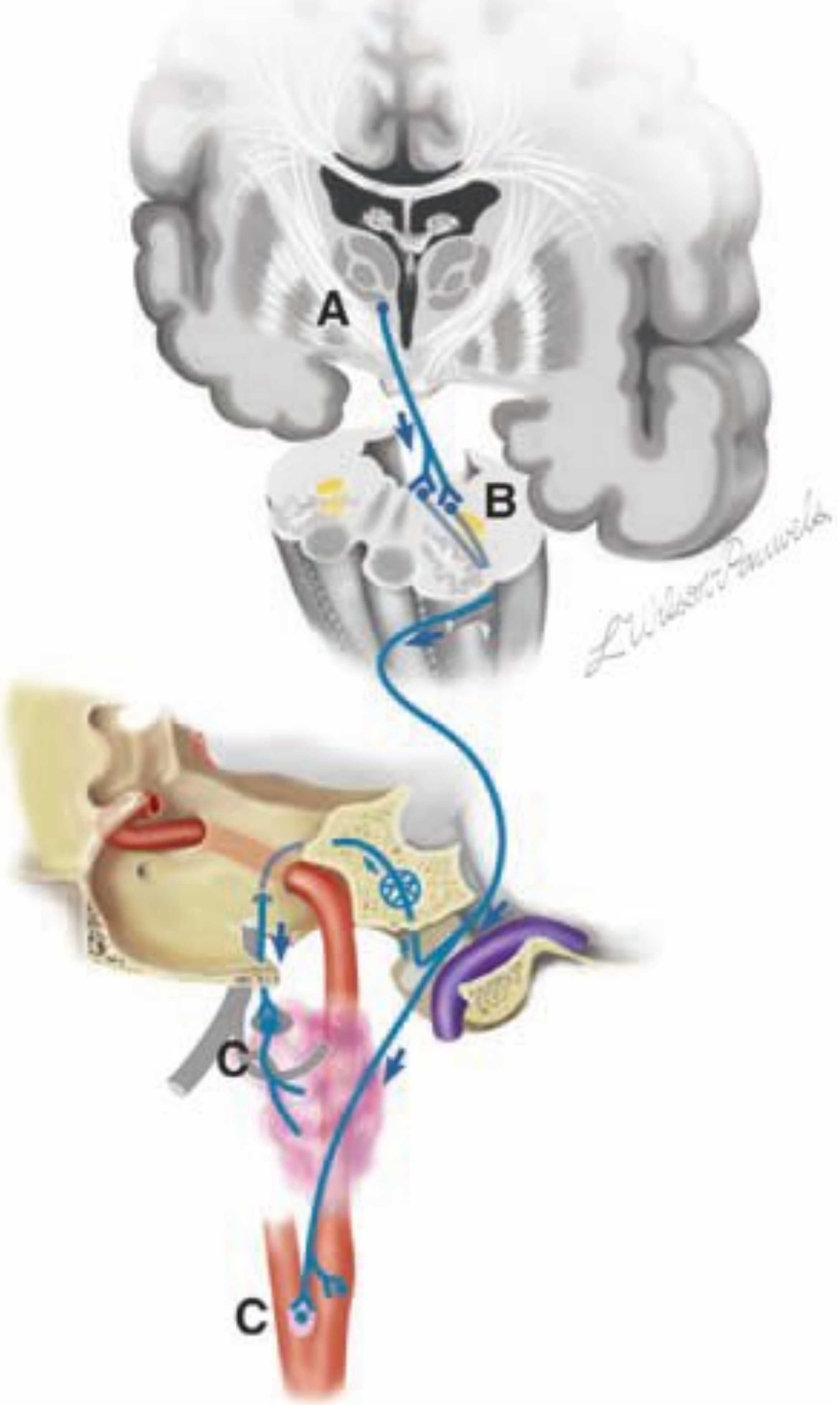
   Damage to any part of the upper motor neuron results in an upper motor neuron lesion (UMNL). The symptoms of an upper motor neuron lesion include

   - Paresis (weakness) or paralysis when voluntary movement is attempted.
   - Increased muscle tone (spasticity).
   - Exaggerated tendon reflexes.

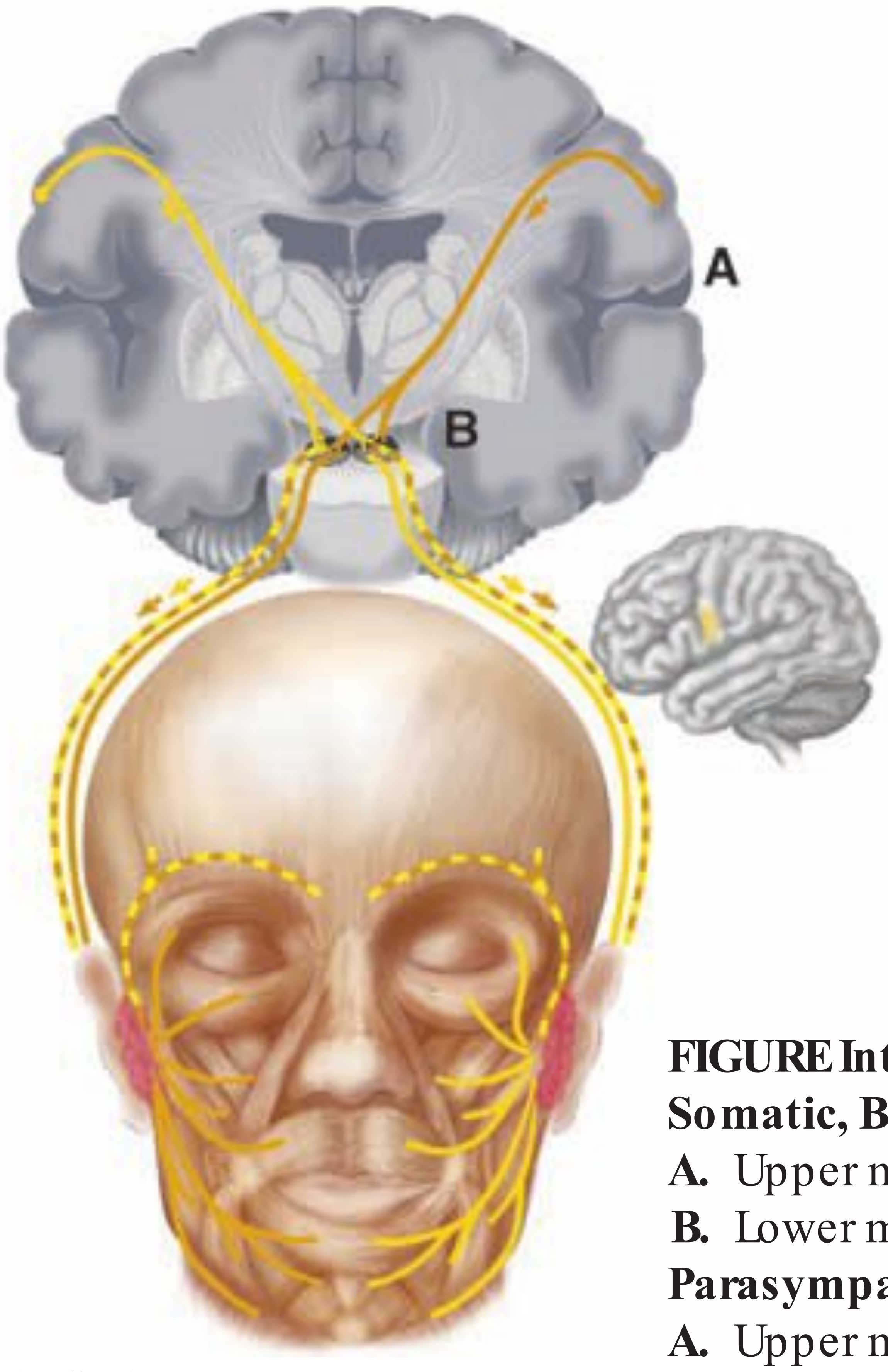
[†] The term "corticobulbar" describes upper motor neuron axons originating in the cortex and terminating in nuclei in the brain stem (bulb).



SOMATIC MOTOR
(see CN XII-1 - to tongue muscles)



PARASYMPATHETIC (VISCERAL) MOTOR
(see CN IX-11 - to parotid gland and carotid body and sinus)



BRANCHIAL MOTOR
(see CN VII-13 - to facial muscles)

FIGURE Intro–4 Examples of motor (efferent) pathways:
Somatic, Branchial
A. Upper motor neuron
B. Lower motor neuron
Parasympathetic
A. Upper motor neuron
B. Lower motor neuron
C. Tertiary cell

Wasting of the muscles does not occur unless the paralysis is present for some time, at which point, some degree of disuse atrophy appears. These symptoms do not occur in those parts of the body that are bilaterally represented in the cortex. In the head and neck, all the muscles are bilaterally represented except the sternomastoid, the trapezius, those muscles below the eyebrow, and the tongue.

2. The **lower motor neuron** is located in the brain stem (Fig. Intro–5) or upper cervical spinal cord. The cell bodies form the motor cranial nerve nuclei. Axons that leave these nuclei make up the motor component of the cranial nerves.

   Damage to any part of the lower motor neuron results in a lower motor neuron lesion (LMNL). The symptoms of a lower motor neuron lesion include

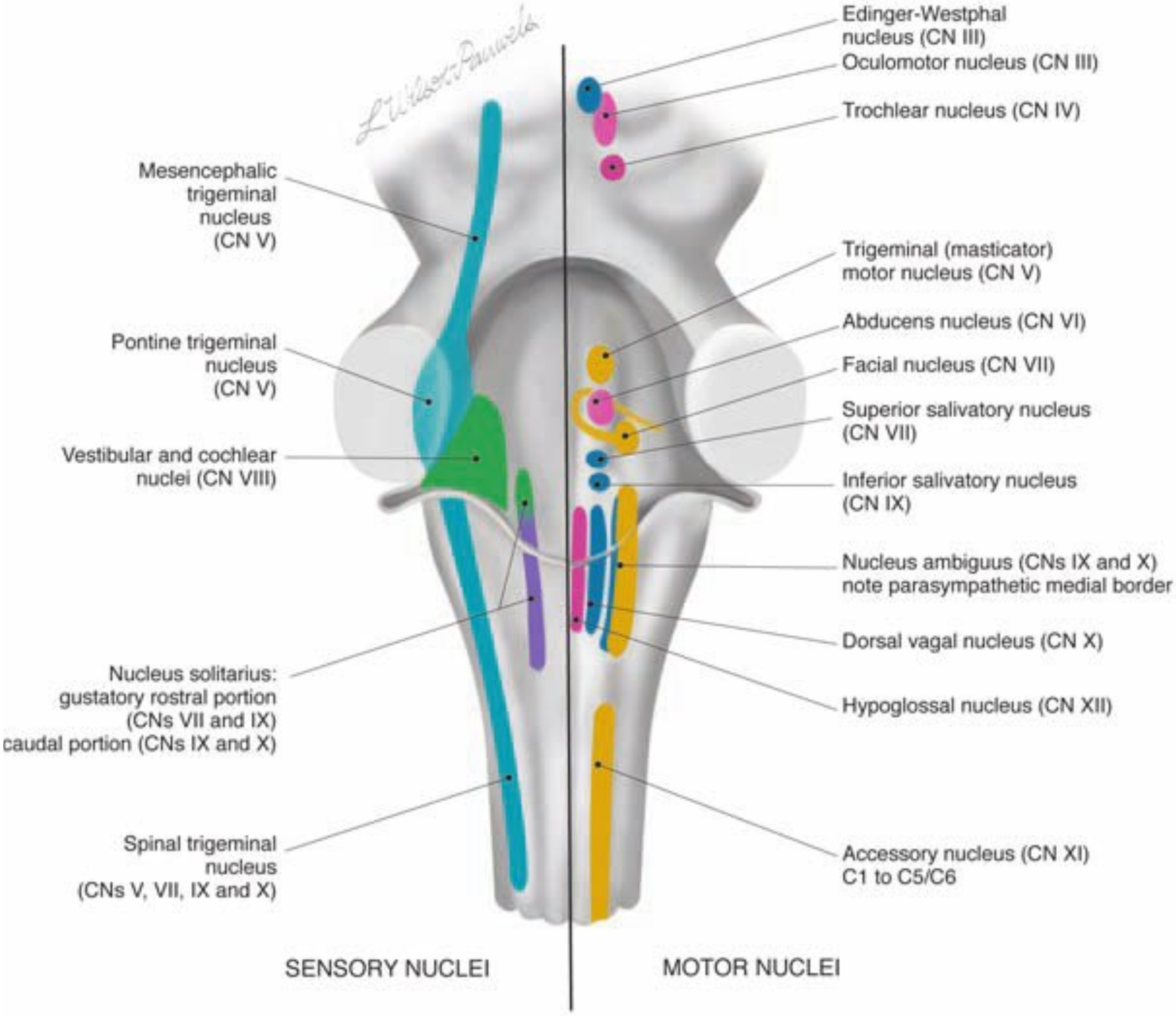

L Wilson-Pauwels
Edinger-Westphal nucleus (CN III)
Oculomotor nucleus (CN III)
Trochlear nucleus (CN IV)
Mesencephalic trigeminal nucleus (CN V)
Trigeminal (masticator) motor nucleus (CN V)
Abducens nucleus (CN VI)
Pontine trigeminal nucleus (CN V)
Facial nucleus (CN VII)
Superior salivatory nucleus (CN VII)
Vestibular and cochlear nuclei (CN VIII)
Inferior salivatory nucleus (CN IX)
Nucleus ambiguus (CNs IX and X) note parasympathetic medial border
Dorsal vagal nucleus (CN X)
Nucleus solitarius: gustatory rostral portion (CNs VII and IX) caudal portion (CNs IX and X)
Hypoglossal nucleus (CN XII)
Spinal trigeminal nucleus (CNs V, VII, IX and X)
Accessory nucleus (CN XI) C1 to C5/C6
SENSORY NUCLEI
MOTOR NUCLEI

FIGURE Intro–5 Cranial nerve nuclei: sensory nuclei shown on left side, motor nuclei shown on right side of the dorsal brain stem.

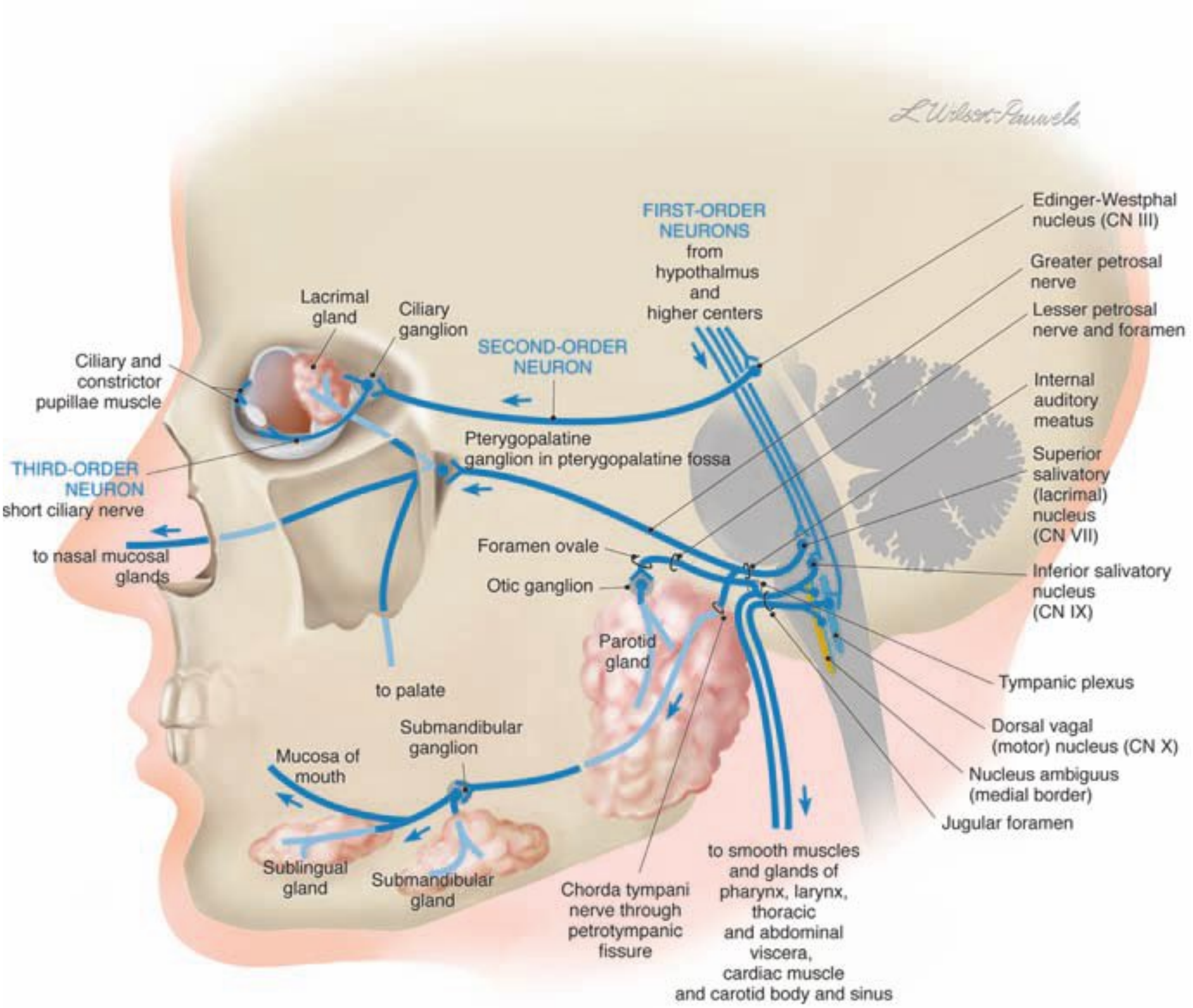
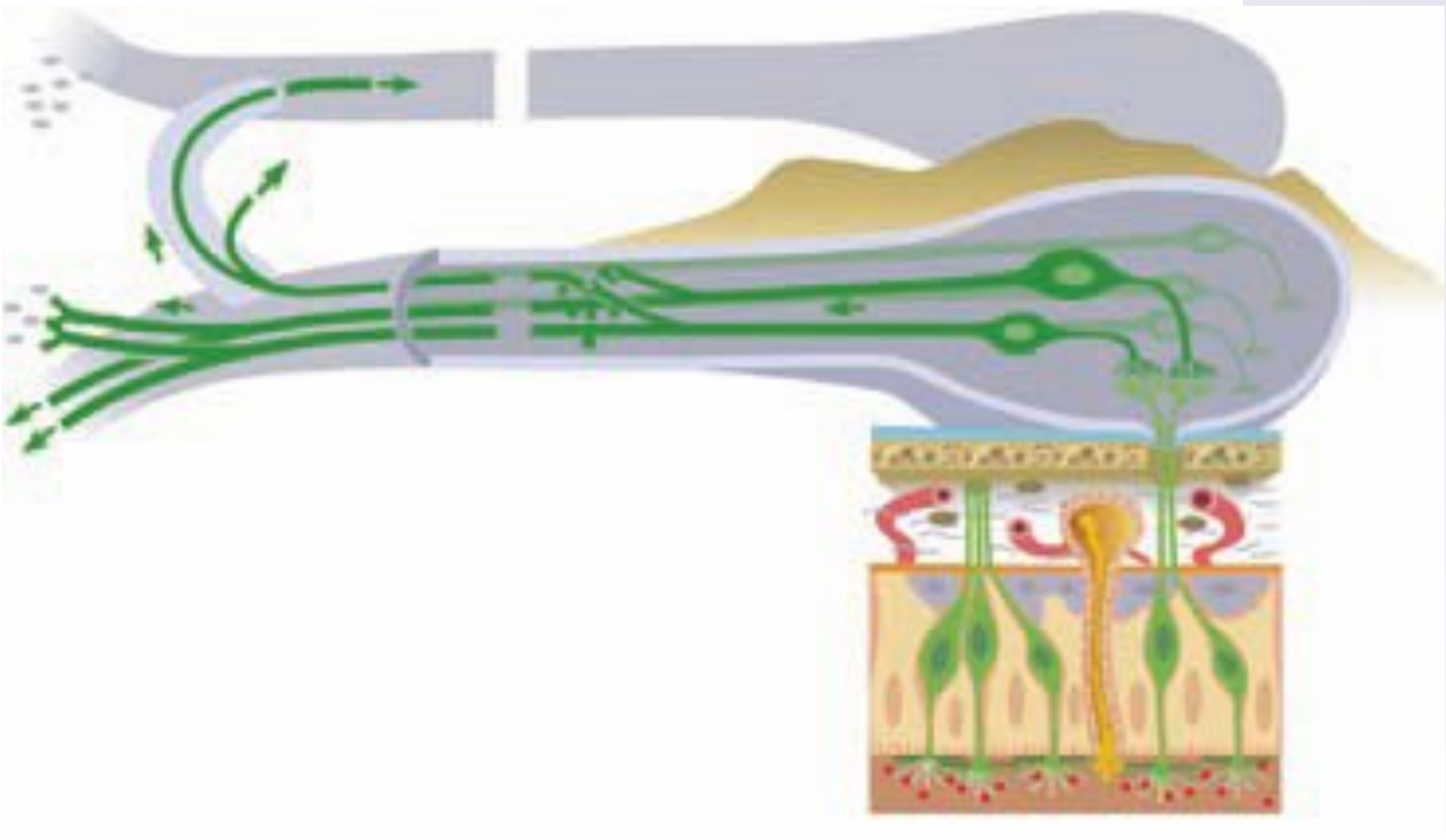

FIRST-ORDER NEURONS
from hypothalmus and higher centers
Edinger-Westphal nucleus (CN III)
Greater petrosal nerve
Lesser petrosal nerve and foramen
Lacrimal gland
Ciliary ganglion
SECOND-ORDER NEURON
Ciliary and constrictor pupillae muscle
Internal auditory meatus
Pterygopalatine ganglion in pterygopalatine fossa
Superior salivatory (lacrimal) nucleus (CN VII)
THIRD-ORDER NEURON
short ciliary nerve
to nasal mucosal glands
Foramen ovale
Otic ganglion
Inferior salivatory nucleus (CN IX)
Parotid gland
to palate
Tympanic plexus
Dorsal vagal (motor) nucleus (CN X)
Submandibular ganglion
Mucosa of mouth
Nucleus ambiguus (medial border)
Jugular foramen
Sublingual gland
Submandibular gland
Chorda tympani nerve through petrotympanic fissure
to smooth muscles and glands of pharynx, larynx, thoracic and abdominal viscera, cardiac muscle and carotid body and sinus

FIGURE Intro–6 Parasympathetic (visceral) motor components of the head and neck.

- Paresis (weakness) or, if all the motor neurons to a particular muscle group are affected, complete paralysis.
- Loss of muscle tone (flaccidity).
- Loss of tendon reflexes.
- Rapid atrophy of the affected muscles.
- Fasciculation (random twitching of small muscle groups).

An example of the parasympathetic (visceral) motor pathway is illustrated in Figures Intro–5 and Intro–6. The involuntary parasympathetic motor pathway differs from the voluntary somatic and branchial motor pathways in that it is a three-neuron chain. Its targets include smooth and cardiac muscle and secretory cells.

1. The **upper motor neuron** is located in a high cortical center. Axons project to parasympathetic nuclei in the brain stem.
2. The **lower motor neuron** is located in the brain stem nuclei and projects via CNs III, VII, IX, and X to ganglia located outside the CNS.
3. The **tertiary neurons** from these ganglia travel to their target cells in the head, thorax, and abdomen (details can be found in each chapter).



I

Olfactory Nerve

CASE HISTORY

Anne, a medical student, was riding to classes when she was knocked off her bicycle at an intersection. She hit the back of her head, suffered a brief loss of consciousness, and was subsequently taken to the hospital. On examination, she was found to have some superficial bruising on her body and her head was tender over the back (occiput) of her skull. Otherwise, she appeared alert and well. A computed tomography (CT) scan of the head (Fig. I–1) revealed a fracture starting at the base of her skull extending through the cribriform plate. Anne was admitted to the hospital for observation overnight.

In the morning, Anne complained that she could not smell anything and, in fact, she could not taste her breakfast. In addition, she had noticed a constant clear discharge from her nose. More complete testing of her cranial nerve functions revealed that her sense of smell was absent (cranial nerve [CN] I). However, direct testing of her taste (CNs VII and IX) continued to be normal. During the next 24 hours, the nasal discharge subsided and eventually Anne was discharged from the hospital.

Five years later, Anne's sense of smell still had not recovered. Her appreciation of food had changed greatly and, although the doctors told her that her taste pathways were intact, food continued to be tasteless.

ANATOMY OF THE OLFACTORY NERVE

The olfactory nerve* functions in the special sense of olfaction or smell, hence, its name (Table I–1). The structures in the central nervous system that are involved in

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*The primary olfactory nerve cell bodies send central processes to synapse on secondary olfactory cells in the olfactory bulb. Secondary sensory axons then form the olfactory tract. Traditionally, the olfactory bulb and tract were known as the olfactory "nerve," but this is no longer the case.

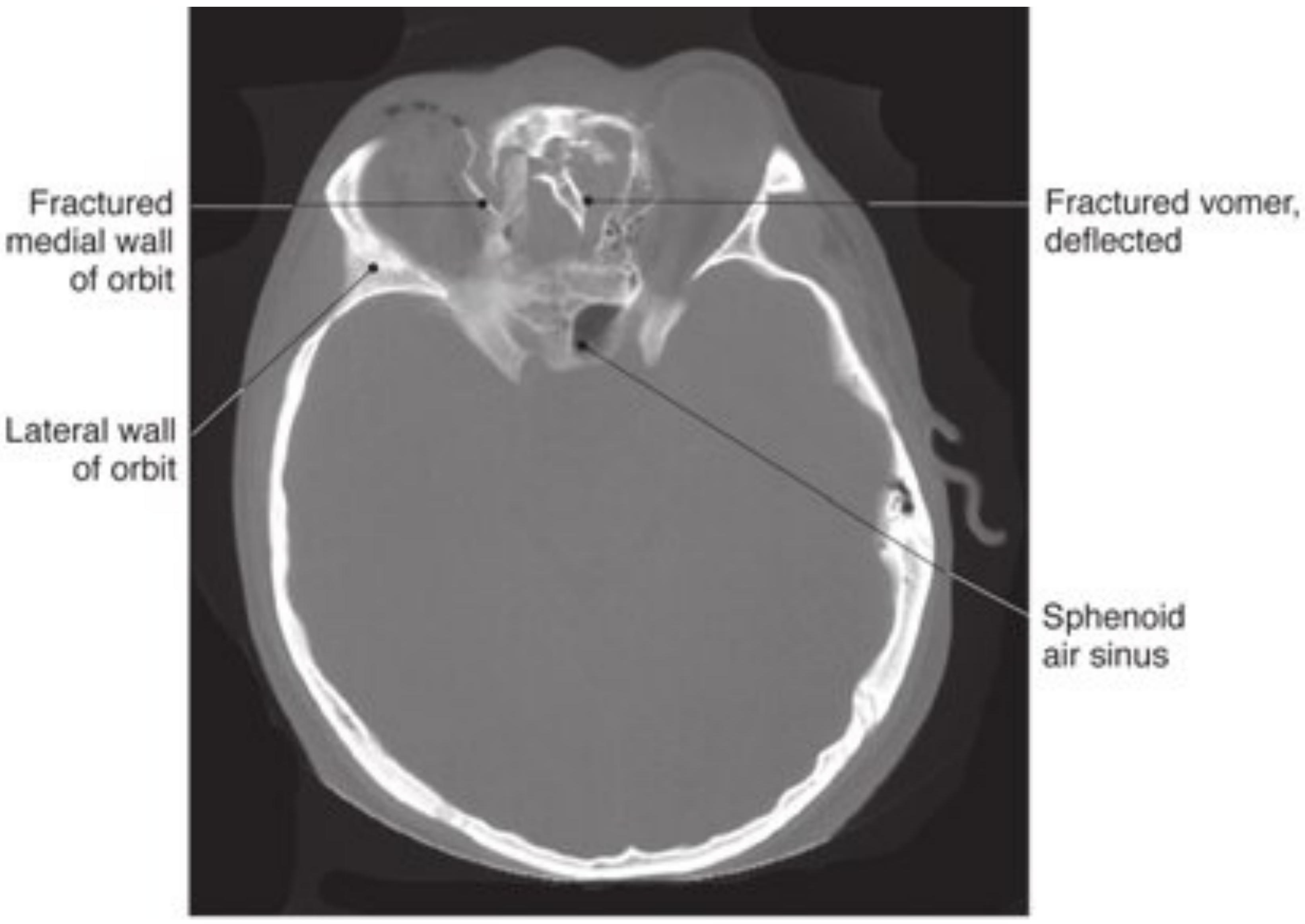

FIGURE I–1 Computed tomography (CT) scan through the vomer showing fractures of the vomer, sphenoid, and medial wall of the orbit. Because the scan is taken in the axial plane, the same plane as the cribriform plate, the fracture of cribriform plate is not visualized in this image. (Courtesy of Dr. Robert Nugent, Department of Radiology, Vancouver General Hospital.)

TABLE I–1 Component, Cells of Origin, and Function of the Olfactory Nerve (CN I)

Component	Cells of Origin	Function
Special sensory (afferent)	Olfactory bulb	For sensation of olfaction or smell

olfaction are collectively called the "rhinencephalon," or "nose" brain. The olfactory system is remarkable in that:

- Peripheral processes of the primary olfactory neurons in the olfactory epithelium (mucosa) (Fig. I–2) act as sensory receptors.
- Primary olfactory neurons undergo continuous replacement throughout life.
- Primary olfactory neurons synapse with secondary olfactory neurons in the olfactory bulb (an outgrowth of cortex), from which signals are sent directly to the cortex without first synapsing in the thalamus. All other sensory pathways synapse in the thalamus before reaching the cortex.
- Pathways to the cortical areas involved in olfaction are entirely ipsilateral.

The olfactory system is made up of the olfactory epithelium, bulbs, and tracts, together with olfactory areas in the brain and their communications with other brain centers.

Olfactory Epithelium

The olfactory epithelium, a specialized area of the nasal mucosa (see Fig. I–2), is located in the roof of the nasal cavity and extends onto the superior nasal conchae

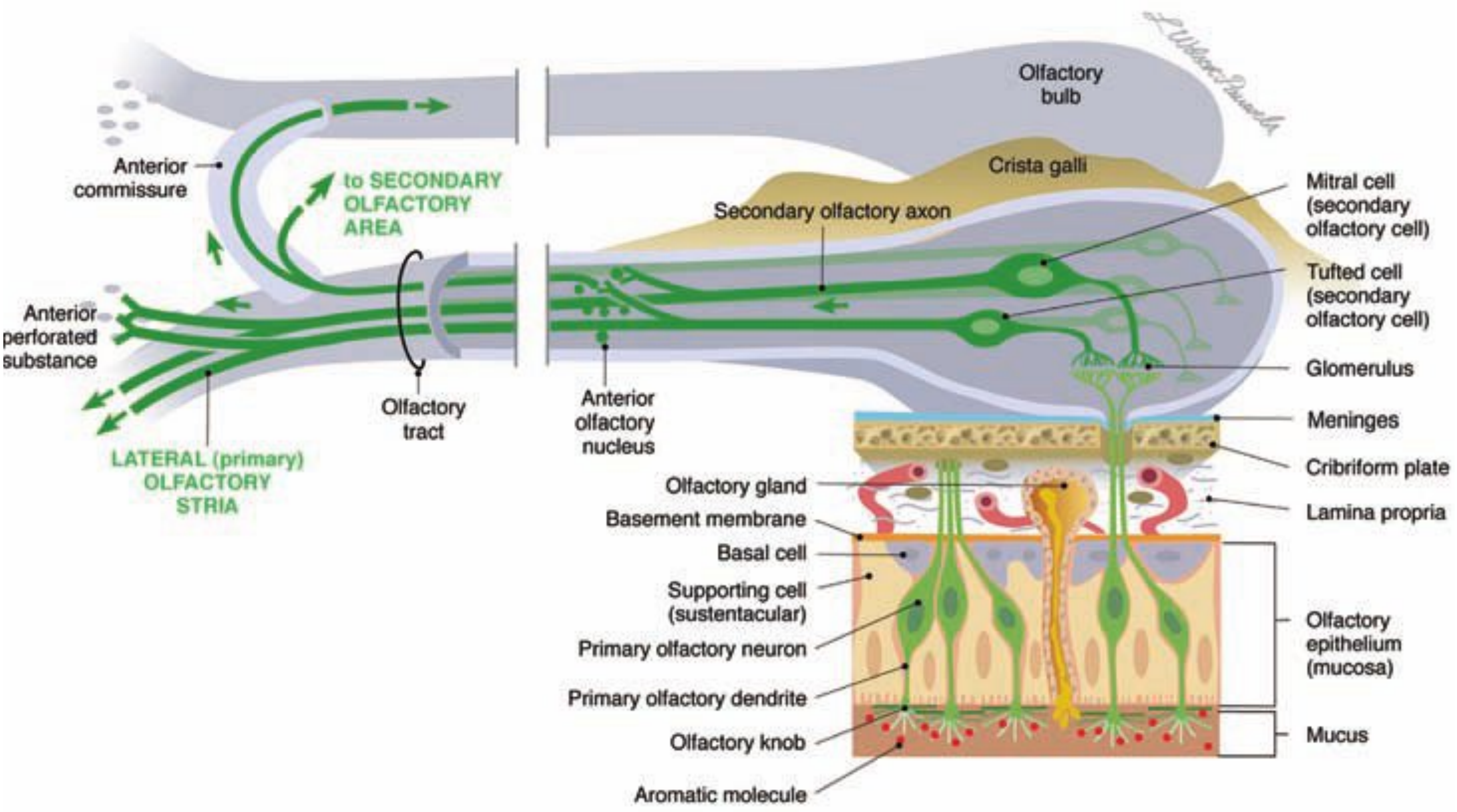
L. Wilson-Pauwels
Olfactory bulb
Crista galli
Anterior commissure
to SECONDARY OLFACTORY AREA
Secondary olfactory axon
Mitral cell (secondary olfactory cell)
Tufted cell (secondary olfactory cell)
Anterior perforated substance
Glomerulus
Olfactory tract
Anterior olfactory nucleus
Meninges
LATERAL (primary) OLFACTORY STRIA
Cribriform plate
Olfactory gland
Lamina propria
Basement membrane
Basal cell
Supporting cell (sustentacular)
Olfactory epithelium (mucosa)
Primary olfactory neuron
Primary olfactory dendrite
Mucus
Olfactory knob
Aromatic molecule

FIGURE I–2 Olfactory epithelium, bulb, and tracts (structures are enlarged for clarity).

laterally and the nasal septum medially. The epithelium is kept moist by olfactory glandular secretions (mucus); it is in this moisture that inhaled scents (odorants) are dissolved. Four principal cell types comprise the epithelium.

1. Primary **olfactory neurons** are bipolar neurons whose peripheral processes (dendrites) extend to the epithelial surface where they expand into olfactory knobs with cilia, which contain the molecular receptor sites. These primary sensory neurons transmit sensation via central processes, which assemble into 20 or more bundles or filaments that traverse the cribriform plate of the ethmoid bone to synapse on the secondary olfactory neurons (mitral and tufted cells) in the olfactory bulb.
2. **Basal cells** lie on the basement membrane and are the source of new receptor cells. This is one of the few types of neurons in the central nervous system that is continuously regenerated throughout life. Regeneration occurs over a period of about 60 days.
3. **Supporting cells** (sustentacular cells) are intermingled with the sensory cells and are similar to glial cells.
4. **Secretory cells** in olfactory glands produce a fluid that includes odorant-binding proteins. The fluid bathes the dendritic endings and cilia of the

receptor cells and acts as a solvent for odorants, allowing their diffusion to the sensory receptors, increasing the efficiency of odor detection.

Olfactory Bulb and Its Projections

The olfactory bulb is the rostral enlargement of the olfactory tract. The olfactory bulbs and tracts are parts of the brain that evaginate from the telencephalon in early development. The olfactory bulb contains the cell bodies of mitral and tufted cells, which are the secondary olfactory neurons involved in the relay of olfactory sensation to the brain. The olfactory bulb also contains spherical structures called "glomeruli" in which contact between the primary olfactory neurons and the secondary olfactory neurons takes place (Fig. I–3). Starting at the cribriform plate, the bulb is arranged in six layers:

1. The nerve fiber layer (olfactory axons) is the most superficial layer, which contains the axons of primary olfactory neurons in the nasal mucosa.
2. The glomerular layer contains spherical glomeruli in which considerable convergence takes place between the axons of primary olfactory neurons and

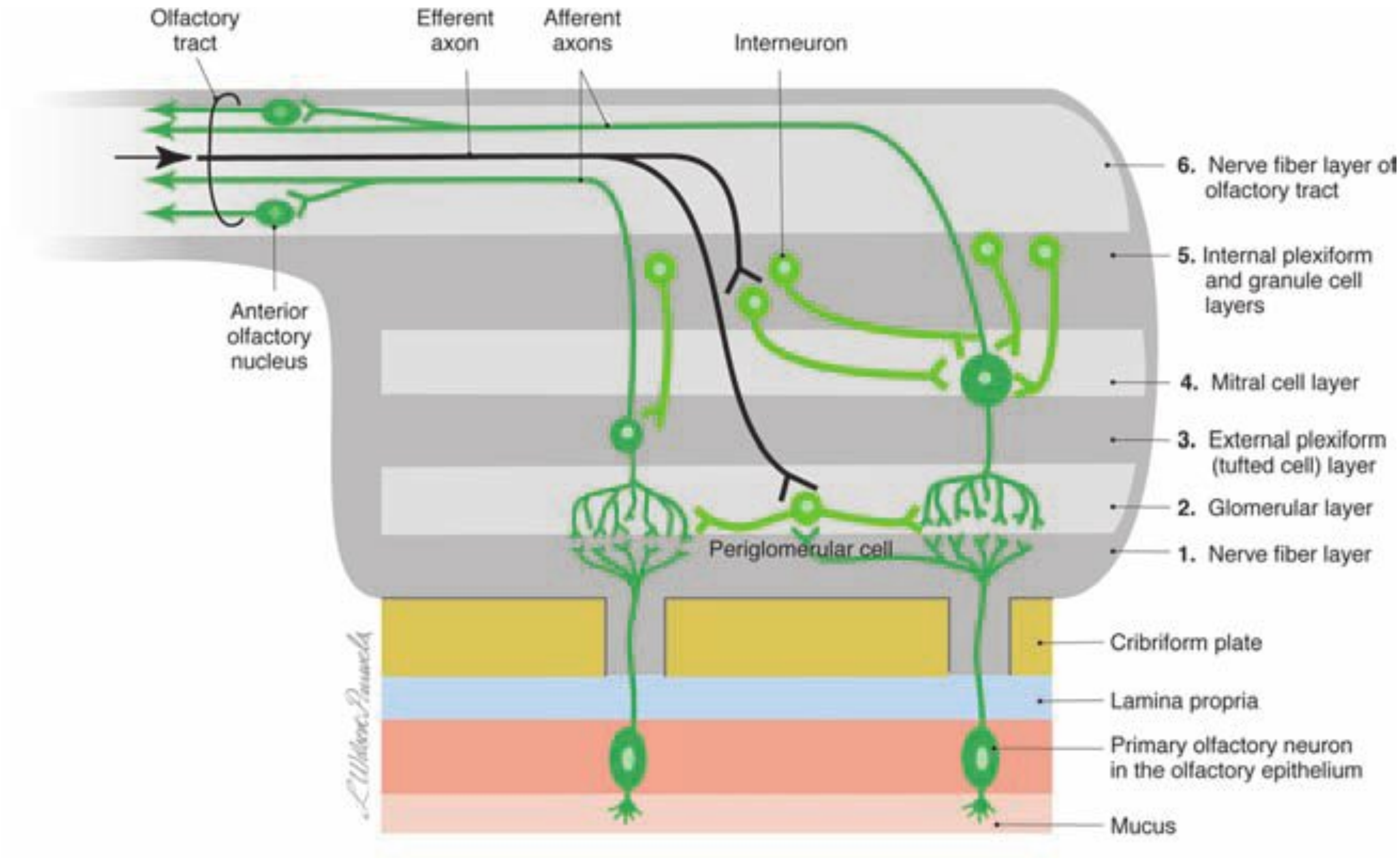
Olfactory tract
Efferent axon
Afferent axons
Interneuron
Anterior olfactory nucleus
6. Nerve fiber layer of olfactory tract
5. Internal plexiform and granule cell layers
4. Mitral cell layer
3. External plexiform (tufted cell) layer
2. Glomerular layer
1. Nerve fiber layer
Periglomerular cell
Cribriform plate
Lamina propria
Primary olfactory neuron in the olfactory epithelium
Mucus

FIGURE I–3 Schematic olfactory pathway from olfactory epithelium to the olfactory tract. Numbers 1 to 6 represent the layers of the olfactory bulb. The olfactory tract includes secondary olfactory axons of tufted and mitral cells; neurons of the anterior olfactory nucleus; and efferent axons from the olfactory cortex and from the contralateral olfactory nucleus.

the dendrites of tufted and mitral cells as well as input from interneurons and motor axons from the olfactory cortex and from the contralateral olfactory nucleus.

3. The external plexiform (tufted cell) layer contains mainly the cell bodies of tufted cells.
4. The mitral cell layer is a single layer of large mitral cell bodies.
5. The internal plexiform and granule cell layers contain axons and collaterals of mitral and tufted cells and a few granule cells.
6. The nerve fiber layer contains myelinated axons of the secondary neurons and motor axons from the cortex.

Within the layers are three major cell types: mitral and tufted cells and the interneuron cells. The mitral and tufted cells are functionally similar and together constitute the sensory neurons from the olfactory bulb to the central nervous system (CNS; see Fig. I–2); the interneurons are made up of periglomerular and granule cells:

1. Mitral cell dendrites extend into the glomeruli where they are contacted by the axons of primary olfactory neurons and by interneurons. After giving off collaterals to the anterior olfactory nucleus, the mitral cell axons project mainly to the lateral (primary) olfactory area.
2. Tufted cell dendrites extend into the glomeruli where they also make contact with the axons of primary olfactory neurons. Their axons project to the anterior olfactory nucleus and to the primary olfactory areas and the anterior perforated substance.
3. Periglomerular and granule interneuron dendrites interact between glomeruli.

Postsynaptic fibers of mitral and tufted cells form the olfactory tract and trigone (an expansion of the olfactory tract just rostral to the anterior perforated substance of the brain). These fibers diverge laterally in front of the anterior perforated substance to form the lateral (primary) olfactory stria to the primary olfactory area for the conscious appreciation of smell (Figs. I–4 and I–5; see also Fig. I–2).[†]

Most of the axons from the olfactory tract pass via the lateral (primary) olfactory stria to the primary olfactory area. The olfactory area consists of the cortices of the uncus and entorhinal area (anterior part of the parahippocampal gyrus), the limen insula (the point of junction between the cortex of the insula and the cortex of the

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[†] A ridge of tissue extending medially from the lateral olfactory area is sometimes identified as the medial olfactory stria; however, recent evidence suggests that there are very few, if any, olfactory axons within this structure. In this text, we have chosen to call the lateral olfactory stria the “lateral (primary) olfactory stria.”

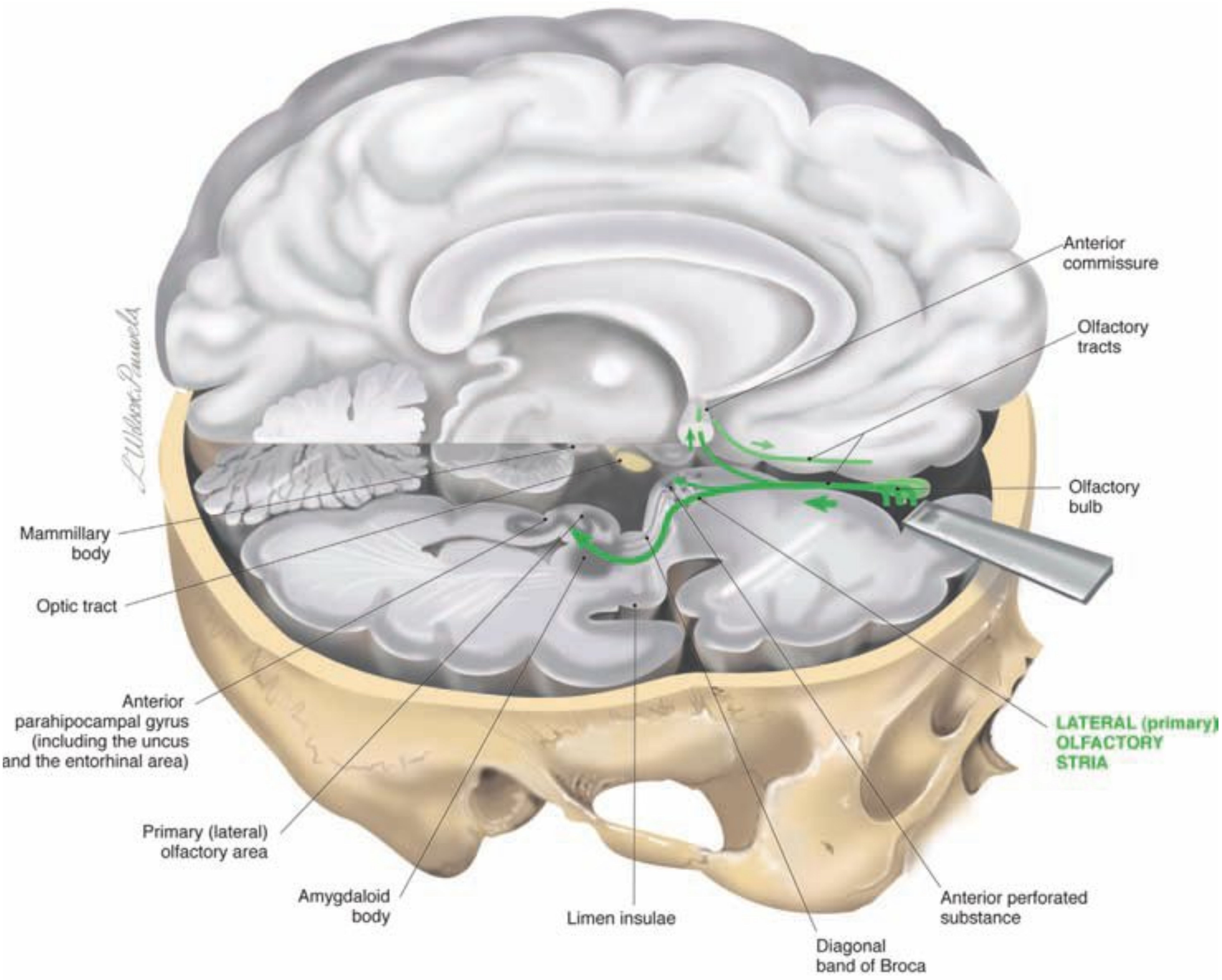
Anterior commissure
Olfactory tracts
Olfactory bulb
LATERAL (primary) OLFACTORY STRIA
Anterior perforated substance
Diagonal band of Broca
Limen insulae
Amygdaloid body
Primary (lateral) olfactory area
Anterior parahipocampal gyrus (including the uncus and the entorhinal area)
Optic tract
Mammillary body

FIGURE I–4 Overview of the olfactory nerve.

frontal lobe), and part of the amygdaloid body (a nuclear complex located above the tip of the inferior horn of the lateral ventricle). The uncus, entorhinal area, and limen insulae are collectively called the "piriform (pear-shaped) area" (see Fig. I–5).

From the primary olfactory areas, profuse projections go directly to the olfactory association area in the entorhinal cortex, sometimes called the "secondary olfactory cortex." It is thought that the primary and secondary olfactory cortices are responsible for the subjective appreciation of olfactory stimuli.

Some collateral branches of the axons of the secondary olfactory neurons terminate in a small group of cells called the "anterior olfactory nucleus," which is a

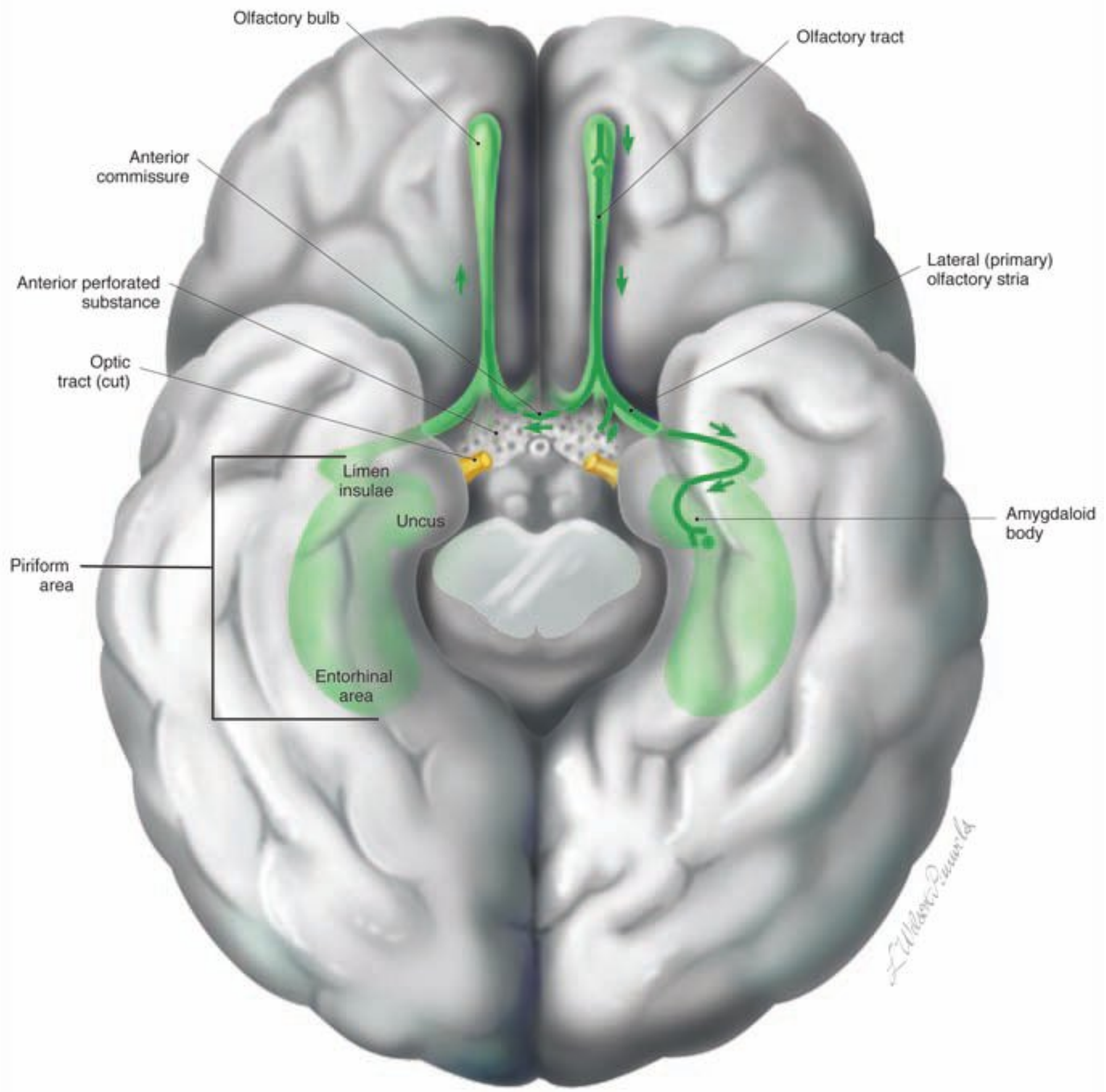

FIGURE I–5 Olfactory areas (inferior view).

collection of nerve cell bodies located along the olfactory tract. Postsynaptic fibers from this nucleus project to the contralateral olfactory bulb via the anterior commissure (see Fig. I–2). Their influence on the contralateral olfactory bulb is mainly inhibitory. This serves to enhance the more active bulb and provide directional cues to the source of the olfactory stimulation.

CASE HISTORY GUIDING QUESTIONS

1. What is anosmia, and why has Anne developed this problem?
2. How is the sense of smell carried from the nasal mucosa to the cerebrum?
3. Where along the olfactory pathway can pathology occur?
4. What is the discharge from Anne's nose?
5. Why is Anne's sense of taste diminished?

1. What is anosmia, and why has Anne developed this problem?

Anosmia is loss of the sense of smell. When Anne fell off her bicycle, she hit the back of her head on the concrete. The blow resulted in an anteroposterior movement of the brain within the cranium. Olfactory axons (filaments) were sheared off at the cribriform plate, resulting in disruption of the olfactory pathway.

2. How is the sense of smell carried from the nasal mucosa to the cerebrum?

Primary olfactory neurons in the nasal mucosa project to the olfactory bulb where they synapse with secondary olfactory neurons. The secondary olfactory cells project centrally via the lateral (primary) olfactory stria to the ipsilateral primary olfactory area in the temporal lobe of the brain (see Figs. I–2 and I–5).

3. Where along the olfactory pathway can pathology occur?

Pathology can occur anywhere along the olfactory pathway. However, it is easiest to consider in terms of the receptor, the primary olfactory axon (filament), and the central pathway.

- *The Receptor*

  Temporary loss of smell results, most commonly, from swelling and congestion of the nasal mucosa due to the common cold or allergic rhinitis, which prevents olfactory stimuli (odorants) from reaching the receptor cells. The receptors themselves can be damaged by chronic smoking and viral infections such as herpes simplex, influenza, and hepatitis. In rare instances, tumors of the olfactory epithelium can arise and are referred to as "esthesioneuroepitheliomas."

- *The Primary Olfactory Axon (Filament)*

  The receptor cells (primary olfactory neurons) transmit their information via axons (processes or filaments) that traverse the cribriform plate of the ethmoid bone to synapse on the secondary olfactory neurons in the olfactory bulb. A blow to the head may cause a shift of the brain, resulting in shearing of the delicate filaments as they pass through the bony cribriform plate. This may result in permanent unilateral or bilateral anosmia and is frequently seen with fractures through the cribriform plate.

- *The Central Pathway*

  The central olfactory pathway includes the olfactory bulb, the olfactory tract, and its central projections. The olfactory bulb may become contused or lacerated from head injury. The location of the bulbs and tracts makes them susceptible to compression from olfactory groove meningiomas, aneurysms of the anterior cerebral artery or the anterior communicating artery, and infiltrating tumors of the frontal lobe. Such compression injuries may result in either unilateral or bilateral loss of smell.

4. What is the discharge from Anne's nose?

Anne's closed head injury resulted in a fracture of the cribriform plate with a subsequent tearing of the dura mater and arachnoid mater. The discharge was cerebrospinal fluid (CSF) leaking through the dural tear into the nasal cavities. The leakage of CSF from the nose is referred to as "CSF rhinorrhea" (Fig. I–6).

The CSF discharge is clear and does not cause irritation within or outside the nose. The patient describes it as "salty-tasting."

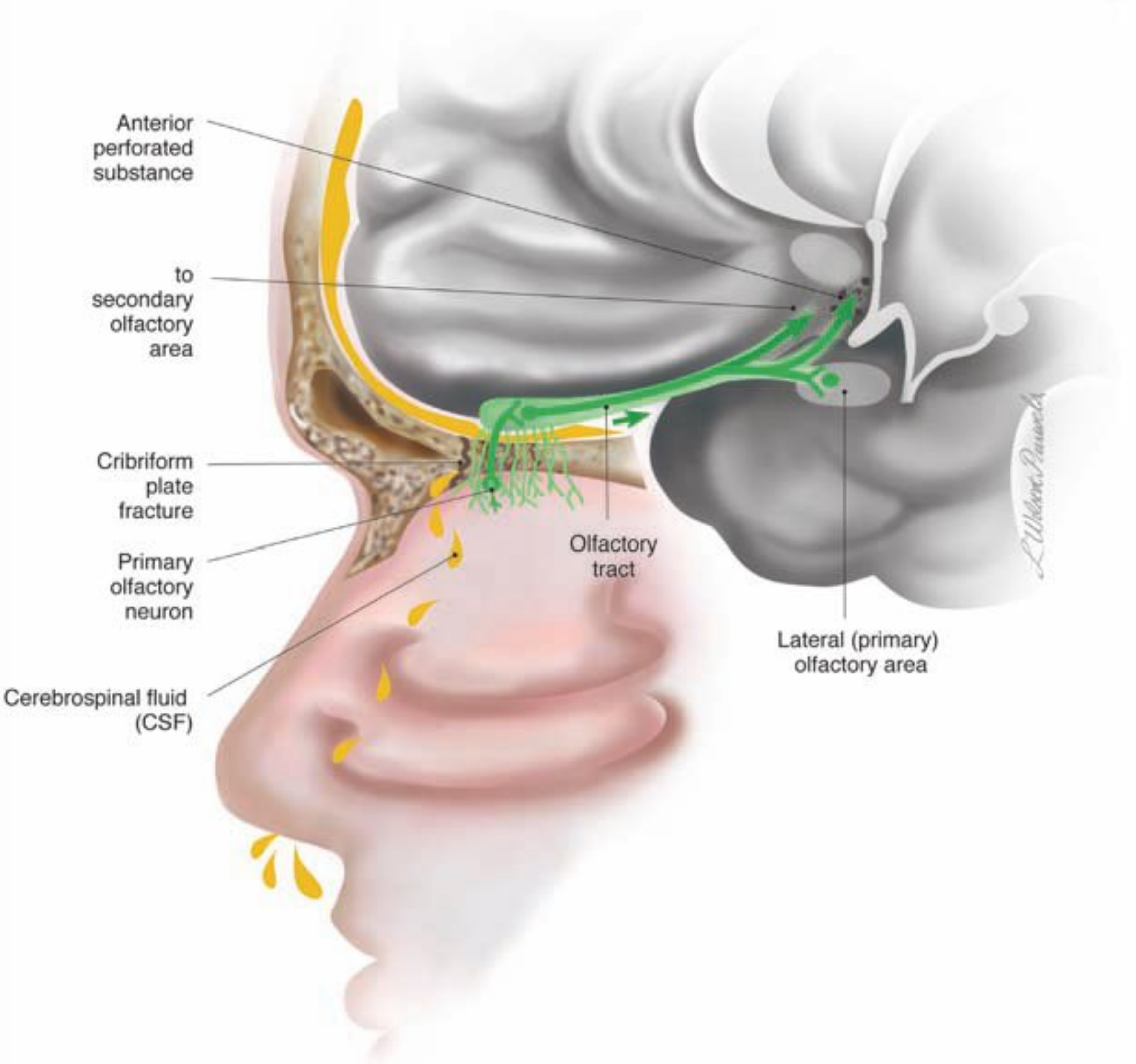
Anterior perforated substance
to secondary olfactory area
Cribriform plate fracture
Primary olfactory neuron
Cerebrospinal fluid (CSF)
Olfactory tract
Lateral (primary) olfactory area

FIGURE I–6 Cribriform plate fracture resulting in cerebrospinal fluid (CSF) leakage.

5. Why is Anne's sense of taste diminished?

Ageusia is loss of the sense of taste (see CNs VII and IX). Perception of flavor is a combination of smell and taste as well as stored memories of the taste of familiar foods. When one contribution to the flavor experience is lost, the overall effect is diminished taste.

CLINICAL TESTING

The olfactory nerve is frequently overlooked in the neurologic examination. However, identification of anosmia is a helpful localizing neurologic finding. The examiner easily can assess the olfactory nerve in the clinical setting by asking the patient to close his or her eyes while the examiner presents a series of nonirritating, familiar olfactory stimuli such as coffee or chocolate (Fig. I–7). The aromatic stimulus should be placed under one nostril while the other nostril is occluded. The patient is asked to sniff the substance and then identify it. The procedure is repeated for the other nostril (see video on the "Cranial Nerves" website). If the patient can name or describe the substance, it is assumed that the olfactory tract is intact. Stimuli such as ammonia are unsuitable because they have an irritative effect on the free nerve endings in the nasal mucosa.

There are more elaborate tests, such as olfactory evoked potentials, that can be used for assessing the olfactory pathway; however, these are primarily research tools and are not used in clinical practice.

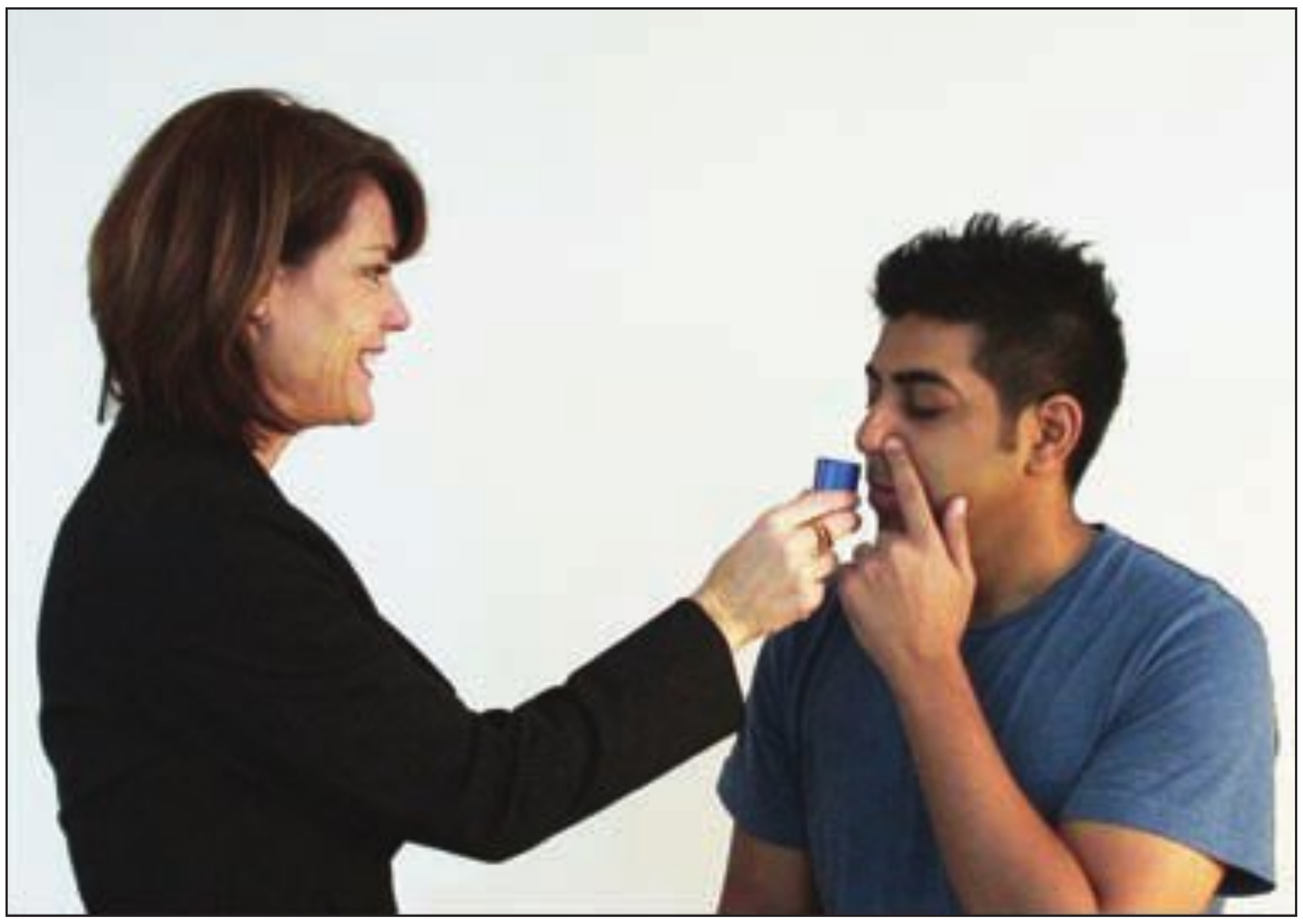

FIGURE I–7 Identifying olfactory stimuli to test for anosmia.

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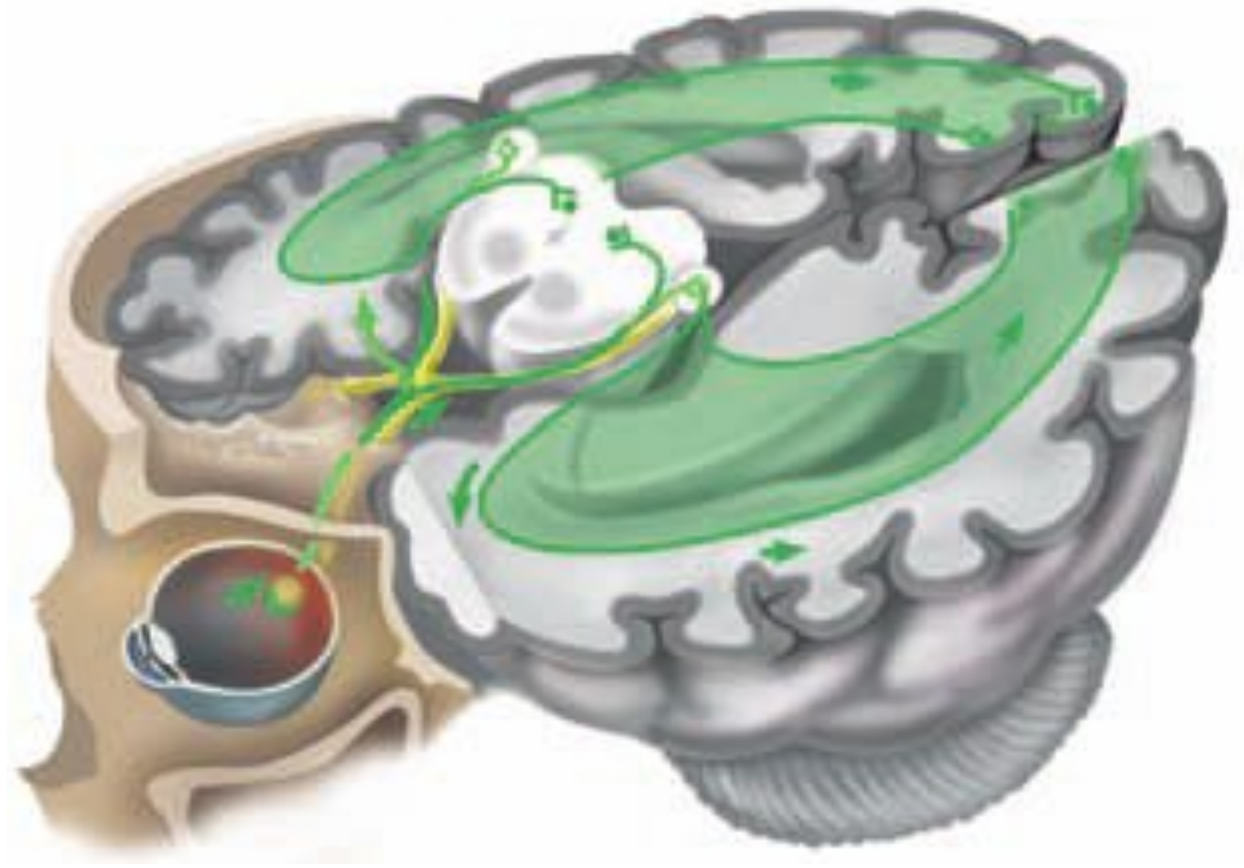

II

Optic Nerve

CASE HISTORY

During a photography session, Meredith, a 28-year-old photojournalist, noticed her vision was blurred in her right eye. Initially, she thought it was the camera lens, but soon realized the problem was her own eye. Her vision continued to deteriorate over the course of the day. When Meredith awoke the next morning, her vision was markedly blurred and her right eye was now painful, especially when she moved it. Quite alarmed by her symptoms, she went to the emergency department of her local hospital. The attending physician wanted to know about her background and whether she had experienced visual loss or any transient motor or sensory symptoms in the past.

Meredith recalled that she had experienced a minor episode of blurred vision in her left eye about a year previously, but it had not interfered with her activities and got better after 1 week. She did not see a doctor at the time because she was at the summer cottage and thought that the symptoms were minor.

The doctor took Meredith's history and then examined her. He noticed that, although her optic disks appeared normal, her visual acuity was decreased to 20/70 in the right eye, whereas in her left eye, it was 20/30. He assessed her color vision using Ishihara plates and found that she had reduced color discrimination, in particular reds appeared less intense, as well as decreased contrast sensitivity in her right eye. When he examined Meredith's visual field, he found that she had a central scotoma (blind area) in her right eye. Both of Meredith's pupils measured 4 mm in diameter. When the doctor shone a bright light into her left eye, both pupils constricted normally; however, when the light was shone in her right eye, there was only a slight constriction of the right and left pupils. The doctor then completed the neurologic examination and found that she had no other neurologic abnormalities at this time.

The doctor told Meredith that she had optic neuritis involving her right eye. He told her that her symptoms might progress further over the next couple of days, but that they should then start to improve.

ANATOMY OF THE OPTIC NERVE

An overview of the optic nerve is illustrated in Figure II–1 and Table II–1. Light enters the eyes and is transformed into electrical signals in the retina. The optic nerve carries these signals to the other parts of the central nervous system. The optic nerve passes posteromedially from the eye to leave the orbit through the optic canal, located in the lesser wing of the sphenoid bone. At the posterior end of the optic canal, the optic nerve enters the middle cranial fossa and joins the optic nerve from the other eye to form the optic chiasma (literally the "optic cross"). A small number of axons from each eye leave the chiasma and course superiorly to the suprachiasmatic nucleus of the hypothalamus where they act to influence the

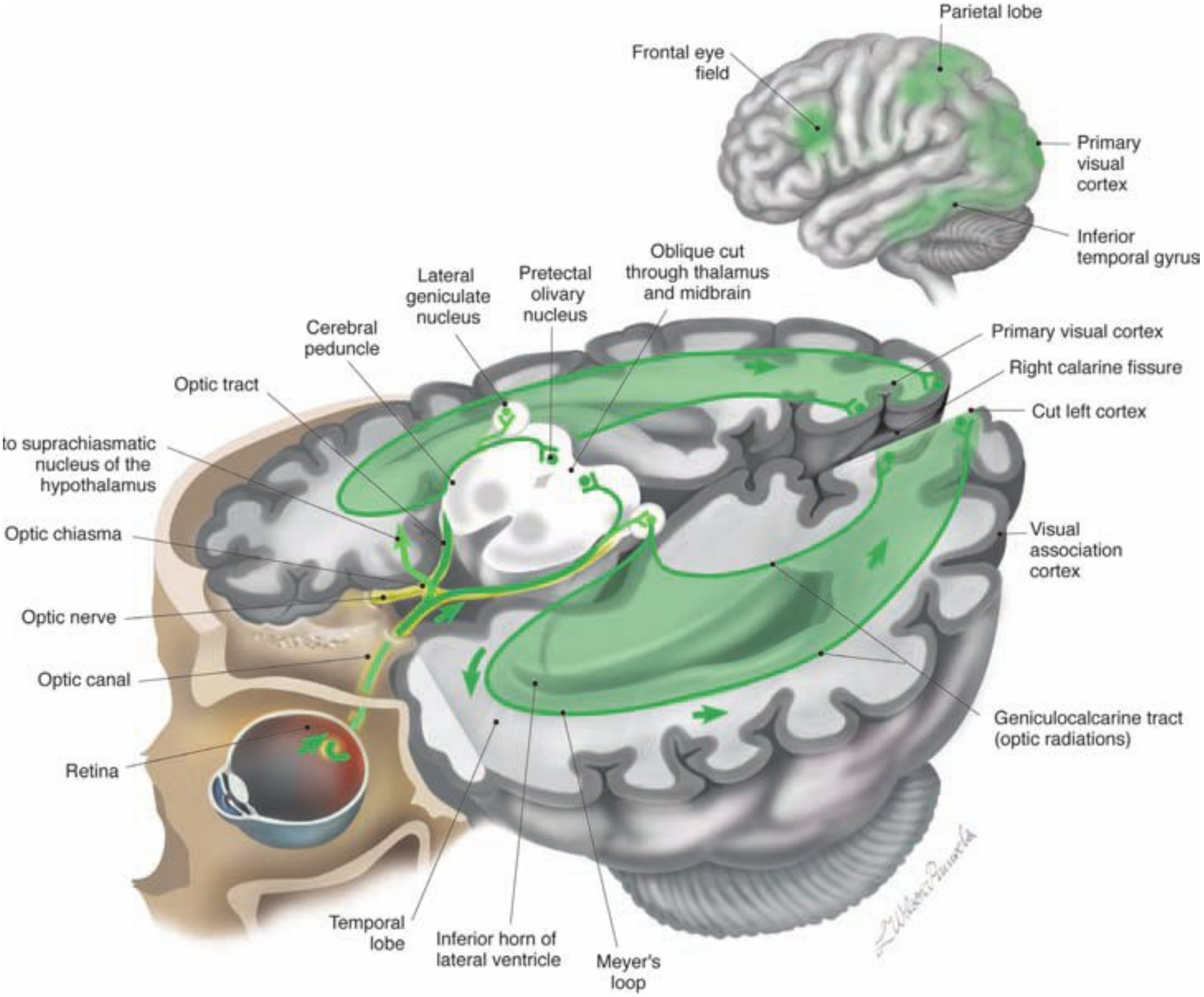
Parietal lobe
Frontal eye field
Primary visual cortex
Inferior temporal gyrus
Oblique cut through thalamus and midbrain
Lateral geniculate nucleus
Pretectal olivary nucleus
Cerebral peduncle
Optic tract
Primary visual cortex
Right calarine fissure
Cut left cortex
to suprachiasmatic nucleus of the hypothalamus
Optic chiasma
Optic nerve
Optic canal
Retina
Visual association cortex
Geniculocalcarine tract (optic radiations)
Temporal lobe
Inferior horn of lateral ventricle
Meyer's loop

FIGURE II–1 Overview of the optic nerve.

TABLE II–1 Component, Cells of Origin and Function of the Optic Nerve (CN II)

Component	Cells of Origin	Function
Special sensory (afferent)	Retinal photoreceptors	For visual information from the retina.

circadian rhythm. At the chiasma, approximately one-half of the axons cross the midline to join the uncrossed axons from the other eye, forming the optic tracts. The optic tracts continue posteriorly around the cerebral peduncles. A small number of the axons leave each tract to terminate in the pretectal olivary nucleus of the midbrain where they form the sensory limb of the pupillary light reflex. A second small group of axons from each tract terminates in the superior colliculus where they orient eye and head movements to a visual stimulus (not illustrated in Fig. II–1). The remaining axons (80%–90%) terminate in the lateral geniculate nucleus of the thalamus (see Fig. II–1).

Axons of lateral geniculate neurons form the geniculocalcarine tract, which is also known as the “optic radiations.” They enter the cerebral hemisphere through the sublenticular part of the internal capsule, fan out above and lateral to the inferior horn of the lateral ventricle, and course posteriorly to terminate in the primary visual cortex surrounding the calcarine fissure. A proportion of these axons form Meyer’s loop by coursing anteriorly toward the pole of the temporal lobe before turning posteriorly (see Fig. II–1).

From the primary visual cortex, integrated visual signals are sent to the adjacent visual association areas and higher-order cortical areas for perception of vision, movement, form, and color and to the frontal eye fields where they direct changes in visual fixation.

Clinical Comment 1

The optic “nerve,” like the olfactory nerve, is not, in fact, a peripheral nerve. In early development, the optic vesicle, an outgrowth of the diencephalon, forms part of the future eye that includes the retina. Axons of retinal ganglion cells then grow back through the stalk of the optic vesicle to contact targets in other parts of the brain, forming the optic nerve, chiasma, and tract. In the strictest sense, the bundle of axons that originate in the retina and terminate in the lateral geniculate nucleus and other areas is a central nervous system tract. Traditionally, however, the part of the tract from the globe to the optic chiasma is known as the “optic nerve.”

The axons in the optic nerve, chiasma, and tract, like other central nervous system axons, are myelinated by oligodendrocytes, not by Schwann cells, which myelinate axons in peripheral nerves. This explains why the optic nerve is affected in multiple sclerosis, a disease of oligodendrocytes, but true peripheral nerves are not.

THE VISUAL PATHWAY

Visual Fields

The visual field is defined as everything we see without moving our eyes or head. The visual field has both binocular (seen with two eyes) and monocular (seen with one eye) areas (Fig. II–2). Light from the binocular area strikes the retina in both eyes. Light from the monocular zone strikes the retina in the ipsilateral eye. Normally, both eyes focus on the same object and view the same visual field but from

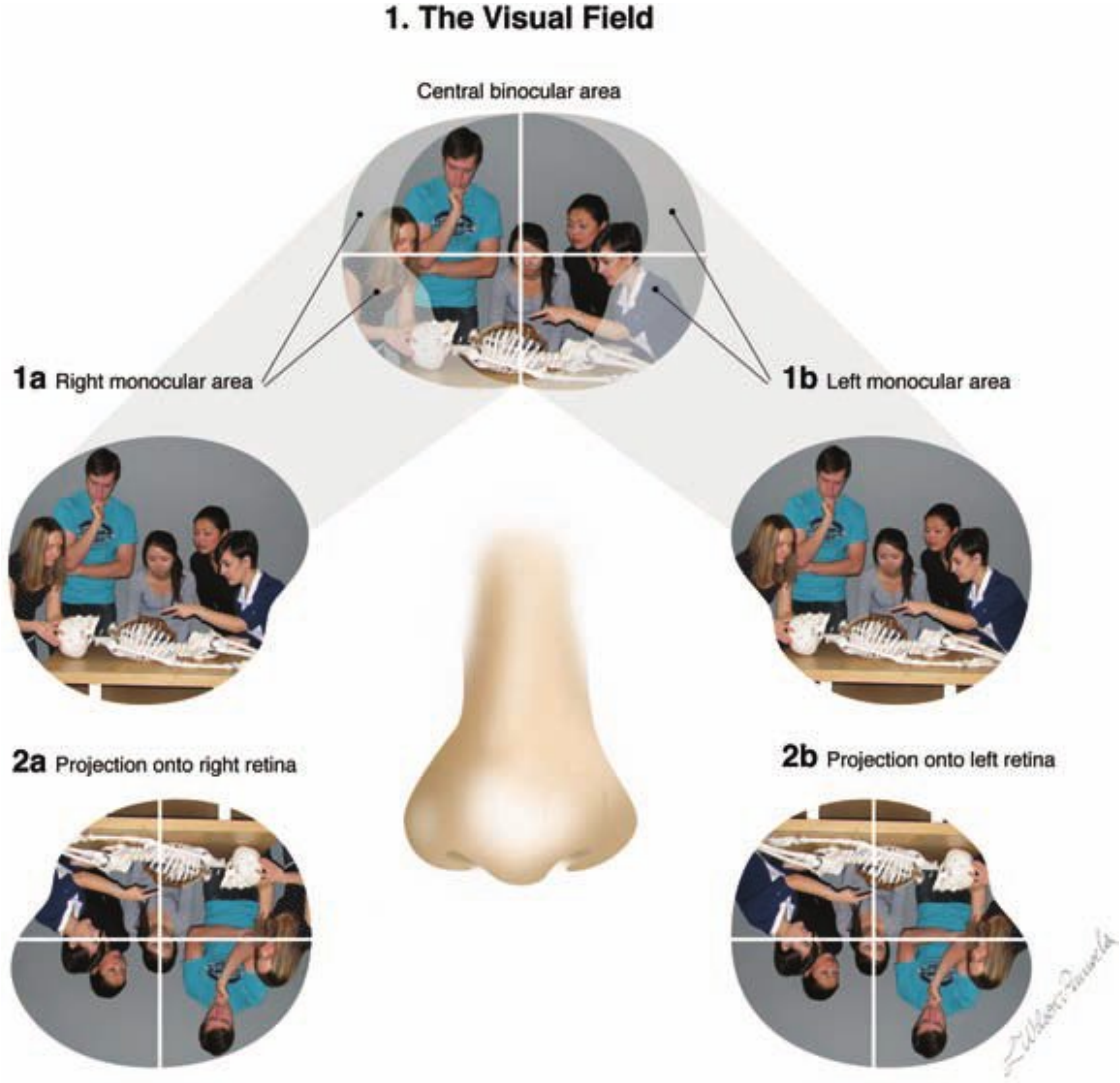

FIGURE II–2 Projection of the visual field onto the retina in each eye. The visual field, **1**, includes a central binocular area, seen by both eyes, and right and left monocular areas seen only by the ipsilateral eye (**1a** &**1b**). The right and left visual fields are projected onto the retinae upside down and reversed (**2a** &**2b**). The center of the visual field, shown by intersecting white lines, is projected onto the centers of the retinae in both eyes (**2a** &**2b**).

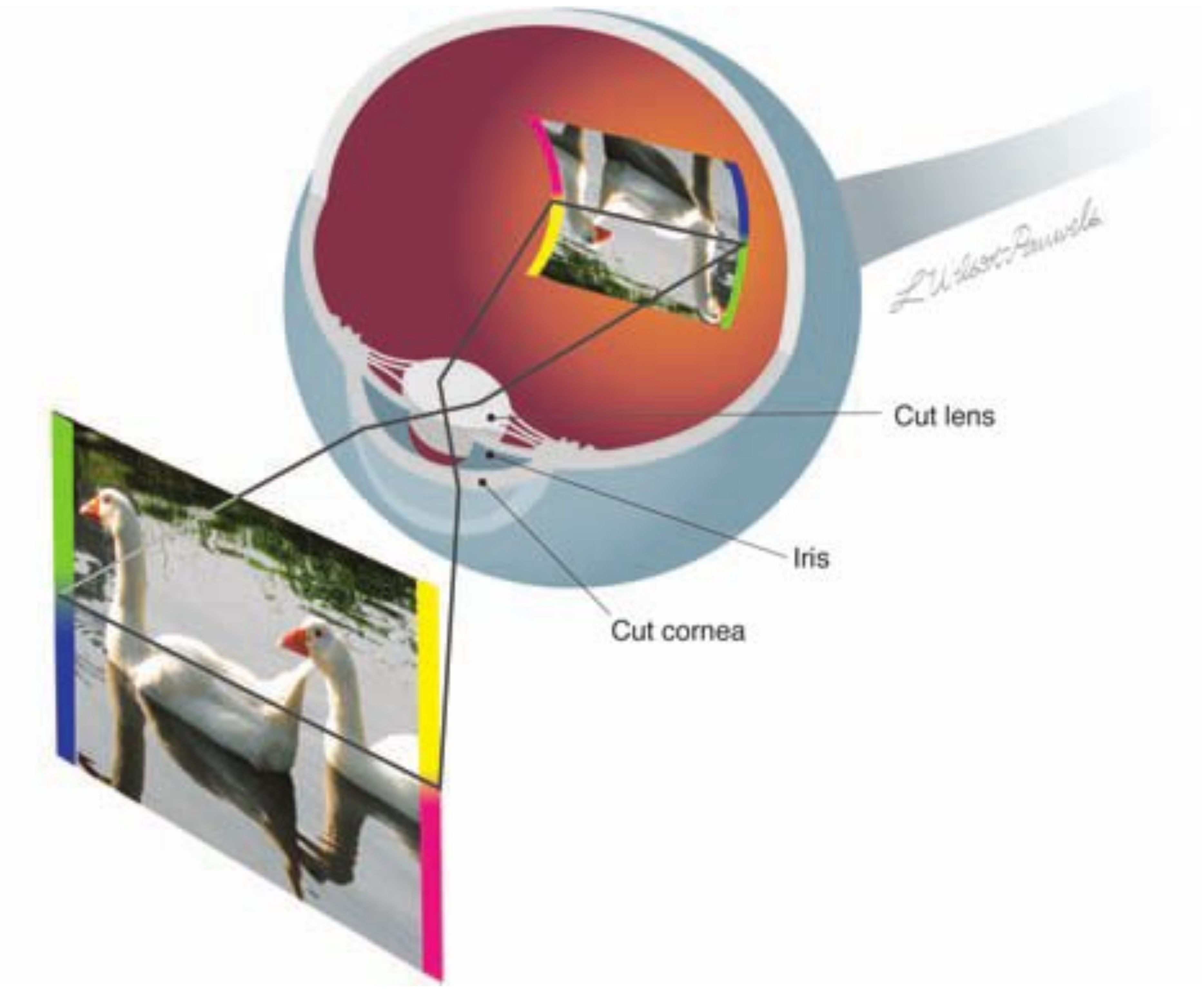

FIGURE II–3 Due to the refraction of light through a convex lens, the visual field is projected onto the retina upside down and laterally reversed.

slightly different angles because of the separation of the eyes. It is this separation that provides stereoscopic vision.

Rays of light from the visual fields are refracted by the cornea and lens before they reach the retina. As a result, the visual fields are projected onto the retinas both upside down and laterally reversed (Fig. II–3).

The Retina

Gross Anatomy

The retina is a specialized sensory structure that lines the posterior half of each eye. It is a cuplike structure whose anterior "rim" is called the "ora serrata." The central point of the retina is indented and is, therefore, called the "fovea (pit)". A small area in (0.35 mm in diameter) in the center of the fovea is called the "foveola" (Fig. II–4A).

An imaginary vertical line through the foveola divides the retina into the nasal hemiretina (close to the nose) and the temporal hemiretina (close to the temporal bone). A second imaginary line, this time in the horizontal plane, further divides the retina into upper and lower halves (see Fig. II–4B). The center of the optic disk lies in the nasal hemiretina just above the horizontal meridian. Optic nerve axons leave the eye, and blood vessels enter the eye at the optic disk. There are no photoreceptors in the optic disk; therefore, it forms a blind spot in the visual field.

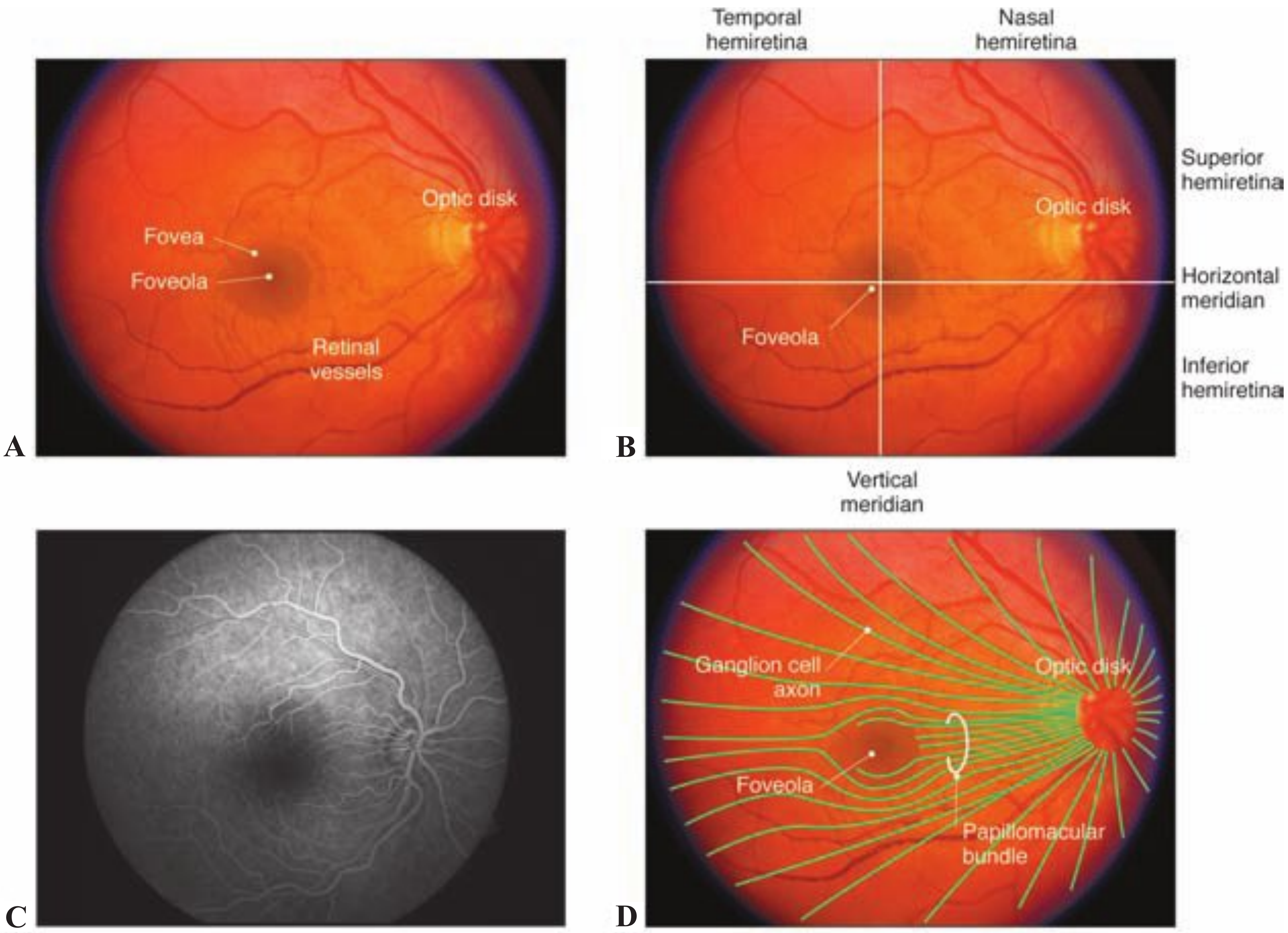

FIGURE II–4 Anatomy of the retina. Right retina shown.

A. Fovea, foveola and the optic disc with emanating retinal blood vessels.
B. The retina is divided into four quadrants centered on the foveola.
C. Retinal angiogram shows that the central fovea is avascular.
D. Ganglion cell axons coursing towards the optic disc, diverting around the fovea and forming the papillomacular bundle.

Retinal fundus photograph courtesy of Dr. R. Buncic, Hospital for Sick Children, Toronto Canada. Retinal angiogram courtesy of Cynthia VanderHoven, BAA CRA, The Hospital for Sick Children.

Light entering the eye travels through the pupil and passes to the back of the retina to reach the photoreceptor layer (rods and cones) where light energy is transduced into electrical signals.* The information received by the photoreceptors is passed forward in the retina to the bipolar cells, which pass the signal further forward to ganglion cells in the anterior layers of the retina. Ganglion cell axons converge toward the optic disk, turn posteriorly, pass through the sclera, and leave the eye as the optic nerve (Fig. II–5). Considerable processing of the retinal signal takes place within the middle layers of the retina.*

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*The details of transduction and of signal processing in the retina are beyond the scope of this text. An excellent description of these processes for beginning scholars can be found in Bear, M.F. Connors, B and Paradiso M. 2007. *Neuroscience: Exploring the Brain*. Chapter Nine.

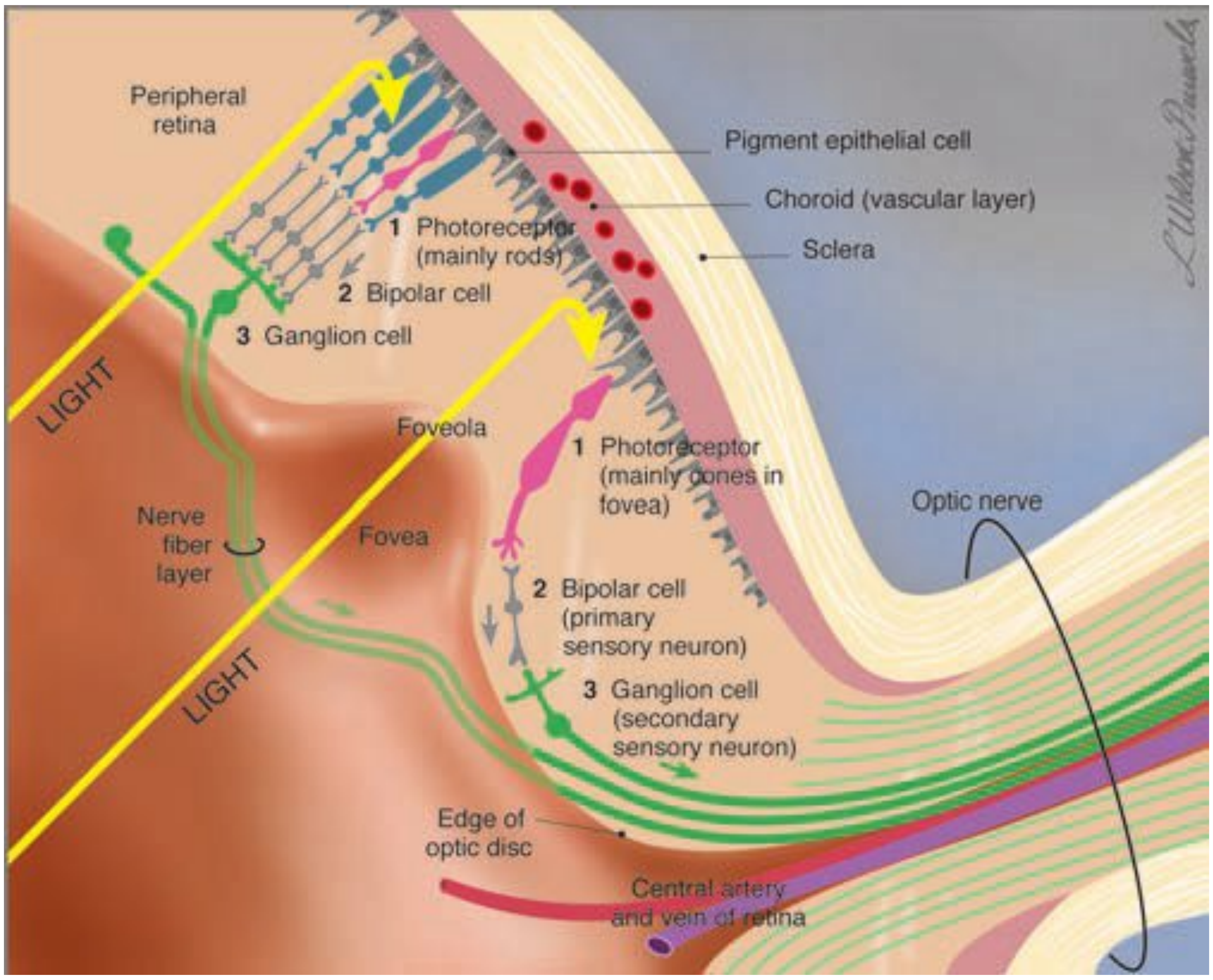

Peripheral retina
Pigment epithelial cell
Choroid (vascular layer)
Sclera
1 Photoreceptor (mainly rods)
2 Bipolar cell
3 Ganglion cell
LIGHT
Foveola
1 Photoreceptor (mainly cones in fovea)
Optic nerve
Nerve fiber layer
Fovea
2 Bipolar cell (primary sensory neuron)
LIGHT
3 Ganglion cell (secondary sensory neuron)
Edge of optic disc
Central artery and vein of retina

FIGURE II–5 Distribution and arrangement of photoreceptors in the central retina compared with the peripheral retina. Cones are the only photoreceptors in the foveola, where they are small, densely packed and have long outer segments. They project to ganglion cells in a one-to-one ratio. In the fovea the inner (anterior) layers of the retina seem to have been pushed aside to allow more direct access of light to the outer segments. The peripheral retina is populated mostly by rods with a small number of cones. Large numbers of photoreceptors project to small numbers of ganglion cells in the peripheral retina.

Clinical Comment 2

Early in development, the distal aspect of the optic vesicle invaginates until it comes in contact with its proximal aspect to form a two-layered optic cup. The outer layer of the optic cup becomes the retinal pigmented epithelium, and the inner layer becomes the remaining nine layers of the retina. The fusion between these layers is not a strong one—it does not include any of the junctional complexes that hold other cell types together—therefore, when the retina detaches, it does so between the pigmented epithelium and the rest of the retina.

Photoreceptors

Photoreceptors are specialized neurons with all the usual cellular components and, in addition, a light-sensitive outer segment composed of stacked layers of

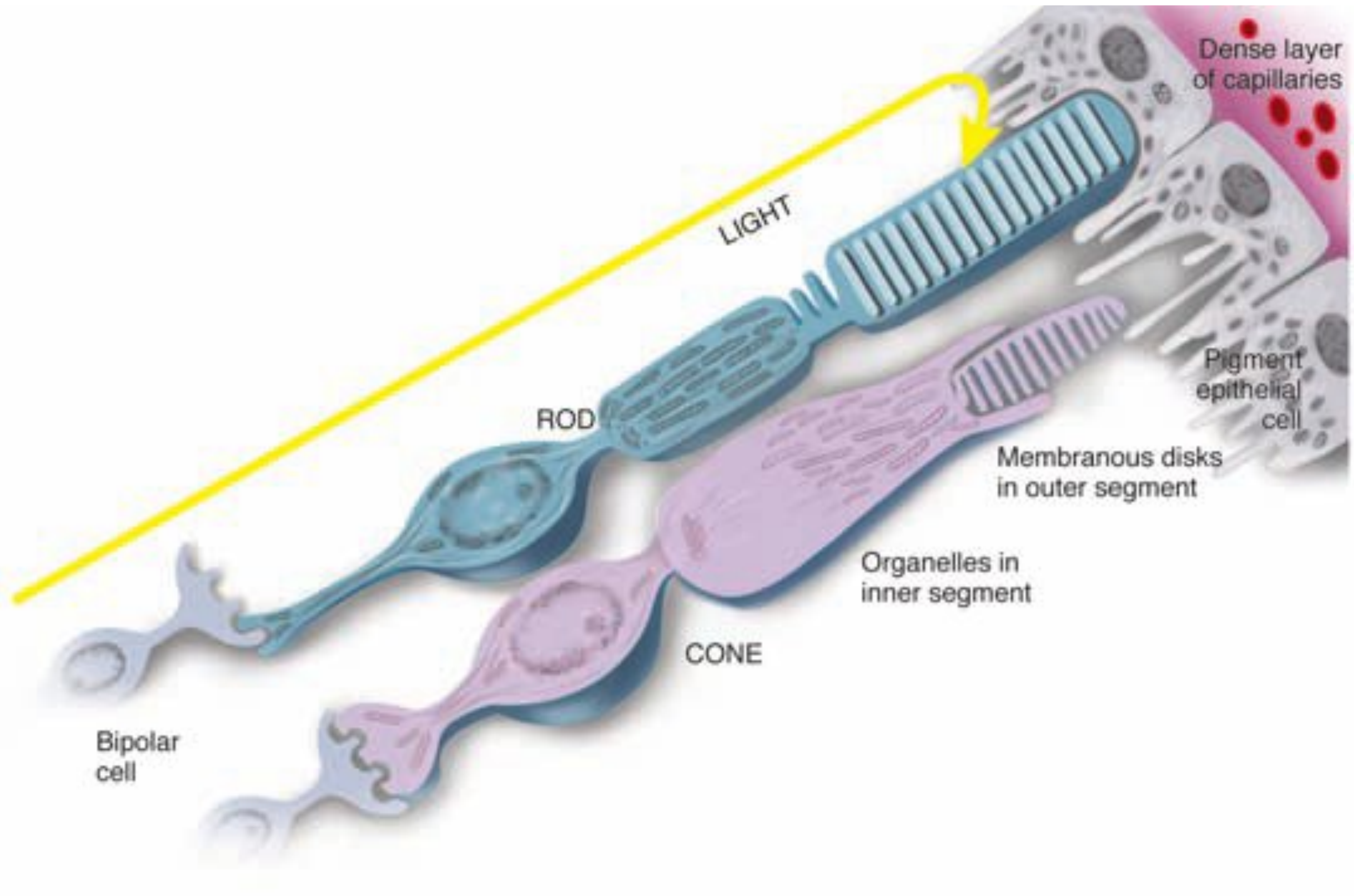

Dense layer of capillaries
LIGHT
ROD
Pigment epithelial cell
Membranous disks in outer segment
Organelles in inner segment
CONE
Bipolar cell

FIGURE II–6 Rods and cones of the retina.

membrane (disks) that contain visual pigments. The disks are continuously produced in the inner segment. Approximately 10% of the disks at the distal end are shed daily and phagocytosed by the pigment epithelial cells; therefore, the outer segment is completely replaced about every 10 days. There are two principal types of photoreceptors: rods and cones (Fig. II–6). A third type of photoreceptor has been described recently. See *Specialist Comment*, later.

Rods function in dim light. They have about 700 disks containing a high concentration of rhodopsin, which makes them highly sensitive to light. They are capable of detecting a single photon; however, they saturate in bright light and are not used in daytime vision. There are approximately 92 million rods in each human retina. They constitute most of the photoreceptors in the peripheral retina and are absent in the foveola. There is a high degree of convergence of rods to ganglion cells and only one kind of rod pigment; therefore, the rod system produces low-resolution, achromatic vision.

Cones function in bright light. The number of disks in their outer segments varies from a few hundred in the peripheral retina to over 1000 in the fovea; however, they have less photopigment than the rods and are, therefore, less sensitive to light. There are approximately 4.6 million cones in the human retina, far fewer than the number of rods. They are present in very low numbers in the peripheral retina, but are found in high densities in the central part of the retina and are the only photoreceptors in the foveola. Cone cells are of three types according to their maximal spectral sensitivities: L, M, and S cones, which absorb light maximally at long (λmax 563 nm), middle (λmax 534 nm), and short (λmax 420 nm) wave lengths in the visible spectrum (sometimes referred to as red, green, and blue cones). They are responsible for color vision. S cones constitute about 10% of cones. They are relatively evenly distributed throughout the retina but are absent from the foveola. Cones are not active in dim light, which is why colors seem to fade at dusk.

Specialist Comment 1

A third, distinct category of photoreceptors has recently been described (Hattar et al., 2006; Berson et al., 2002; Berson, 2003). A small number of retinal ganglion cells are intrinsically photosensitive. They contain melanoposin, a primitive visual pigment, and they are thought to monitor overall levels of illumination in the retina. This non–image-forming pathway is the major route by which the retina influences the circadian rhythm via its target cells in the suprachiasmatic nucleus and the pupillary light reflex via its target cells in the pretectal olivary nuclei.

Clinical Comment 3

The absence of L cones (protanopia, 1% of males, 0.01% of females) or the absence of M cones (deuteranopia, 6% of males, 0.4% of females) results in red-green colorblindness. This condition is sex-linked because the genes for both L and M cones are located on the X chromosome. Absence of S cones (tritanopia, 0.01% both males and females) produces a visual defect in the blue range.

Ganglion Cells

There are approximately 1 million ganglion cells in each human retina. Most of them function in the image-forming pathway. They receive signals from the rod and cone photoreceptors via bipolar cells and send signals to several targets in the central nervous system, principally the lateral geniculate nucleus. Ganglion cell axons carrying signals from the fovea form a distinctive large group of axons, called the "papillomacular bundle," as they course toward the optic disk (see Fig. II–4D). Within the retina, ganglion cell axons are not myelinated, because myelin refracts light, but once they enter the optic nerve, they acquire a myelin sheath, which provides for rapid transmission of the visual signal.

Specialist Comment 2

Several different categories of ganglion cells have been described. Two of them, P cells and M cells, account for 80% of the total. They constitute the image-forming system and they project to the lateral geniculate nucleus. P cells have small receptive fields. They receive signals from single cones in the foveola via midget bipolar cells, providing for the high resolution in central vision. M cells have much larger receptive fields, which is consistent with their presumed role in motion detection. An easy way to remember these two types of ganglion cells is P for particulars and M for movement. The remaining ganglion cells, approximately 200,000, some of which are intrinsically photosensitive, project to other targets in the thalamus, pretectum, and midbrain to drive visual reflexes.

Central Vision

Although we perceive vision as having equally high resolution and color discrimination across the entire field, in fact, they are only high within the central visual field, that is, the fovea and area immediately surrounding it (Fig. II–7). Both resolution and color discrimination are much reduced in parts of the retina outside of the fovea.[†] Central vision is critically important in human vision; without it we cannot read or recognize faces and we are classified as legally blind.

High resolution and color discrimination in central vision is produced by (1) the type and arrangement of photoreceptors in the central retina and (2) anatomic modifications of the fovea.

1. Only cones populate the central retina, and they are responsible for color discrimination. The cones in the fovea are narrow and densely packed, and they have long outer segments compared with those in the peripheral retina. As a result, a single foveolar cone is capable of responding to light from a very small area of the visual field. Each foveolar cone sends signals to a single ganglion cell, giving the cone its own dedicated line to the visual cortex. In contrast, large numbers of photoreceptors in the peripheral retina project to individual ganglion cells, producing a low-resolution image (see Figs. II–5 and II–7). Approximately half of the ganglion cells in each retina receive signals from photoreceptors in the fovea and the area just around it; the remaining half represent the entire peripheral retina. As a result, half of the axons in the optic nerves, chiasma, and tracts, and in fact, half of the entire visual system behind the retina, is concerned with central vision.
2. Because the vertebrate retina is inverted—that is, the photoreceptors are at the back of the retina not at the front where light rays first strike—photons (light energy) must traverse all the cellular layers of the retina, but are blocked by blood vessels that supply it, before they encounter the photoreceptors (see Fig. II–5). The fovea has the following anatomic modifications that facilitate the passage of light to the photoreceptors.

[†] You can prove this to yourself by doing the following experiment. Ask a friend (person A) to stand 15 feet or so in front of you and hold up some fingers. While you are looking directly at person A, her or his image is falling on your foveas. In fact, this visual fixation is called “foveating.” You will have no trouble counting the number of fingers your friend is holding up. Then ask a second person (person B) to stand 5 feet or so to one side of person A and hold up some fingers. As long as you are foveating person A, person B’s image is falling on an area of your retina outside of your fovea. While looking directly at person A, see if you can count the number of fingers person B is holding up. You will not be able to. You will also have trouble seeing what color person B’s clothes are. In fact, you would not be able to identify person B if you did not already know her or his identity.

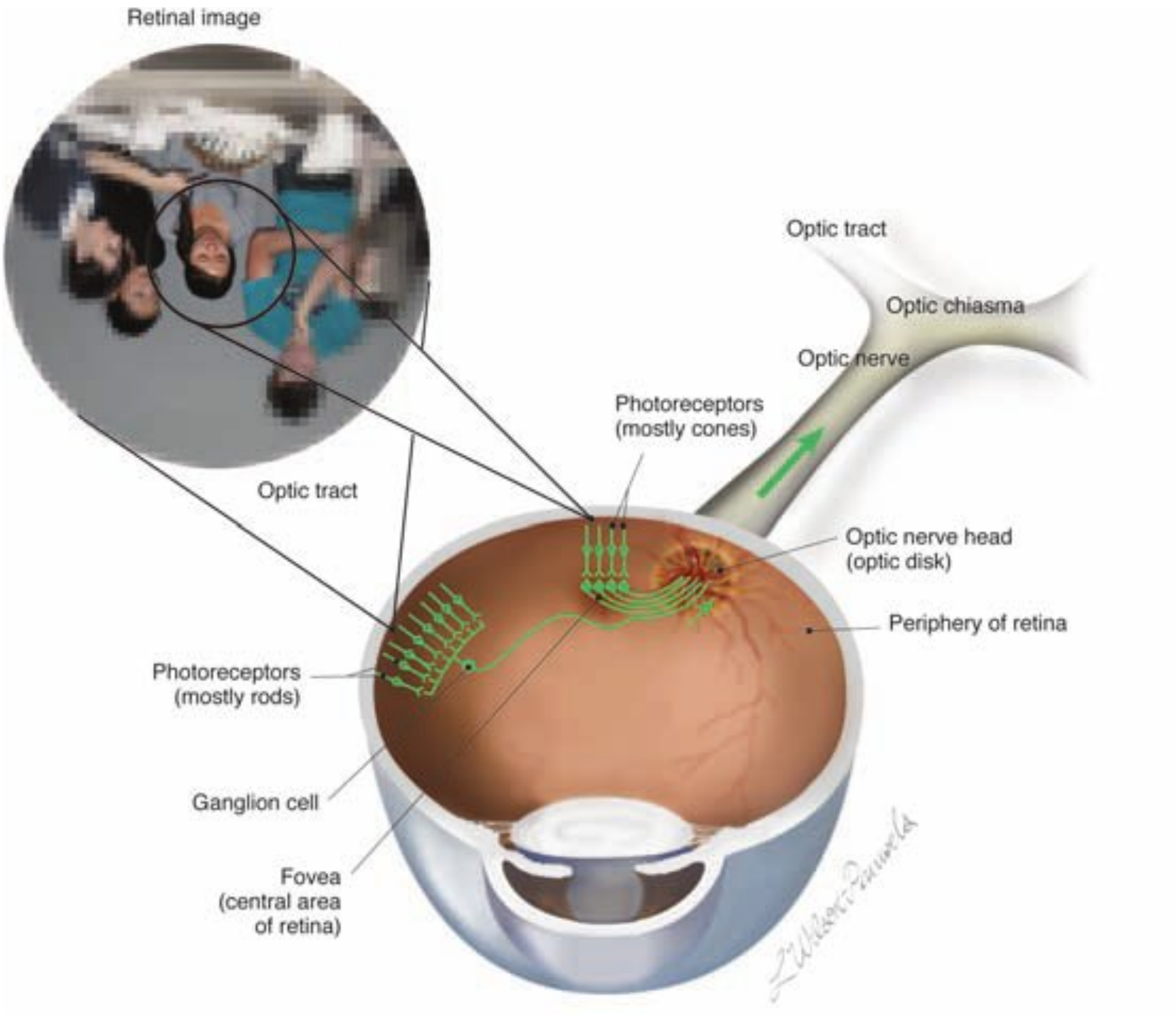

Retinal image
Optic tract
Optic chiasma
Optic nerve
Photoreceptors (mostly cones)
Optic tract
Optic nerve head (optic disk)
Periphery of retina
Photoreceptors (mostly rods)
Ganglion cell
Fovea (central area of retina)

FIGURE II–7 Convergence of photoreceptors on ganglion cells. The retinal image shows high visual resolution and good color discrimination in the center of the visual field. These features are generated by a high cone density and one-to-one projection of cones to ganglion cells. The blurry periphery of the retinal image depicts the low resolution generated by the projection of a large number of photoreceptors, mainly rods, to a small number of ganglion cells. Color discrimination in the peripheral retina is reduced because of the reduced density of cones.

- The fovea is avascular, that is, there are no capillaries in front of the photoreceptors to deflect the light rays (see Fig. II–4C). Instead, the photoreceptors in the fovea are supplied with oxygen and nutrients by a dense bed of capillaries behind the pigmented epithelium (see Fig. II–5).
- Most ganglion cell axons take the most direct path toward the optic disc. However, those whose direct route would take them across the front of the fovea divert around it so as not to interfere with central vision (see Figs. II–4D and II–5).
- In the foveola, the inner (anterior) layers of the retina seem to have been "pushed aside" to allow light direct access to the photoreceptor outer segments (see Fig. II–5).

Optic Nerve, Chiasma, and Tracts

Ganglion cell axons carrying visual information from the four retinal quadrants converge toward the optic disc in an orderly fashion and maintain approximately the same relationship to each other within the optic nerve. Within the chiasma, axons from the nasal halves of both retinas cross the midline. This arrangement of axons results in the information from the right half of the visual field from both eyes being carried in the left optic tract and the left half of the visual field from both eyes being carried in the right optic tract (Fig. II–8).

Lateral Geniculate Nucleus and Geniculocalcarine Tract

The lateral geniculate nucleus of the thalamus is a highly organized structure composed of six layers (Fig. II–9). The two ventral layers are composed of relatively large neurons and are called the "magnocellular layers." They receive signals from the magnocellular ganglion cells, which arise mainly from the peripheral retina and encode movement. The four dorsal layers are composed of smaller neurons and are called the "parvocellular layers." They receive signals from parvocellular ganglion cells, which carry signals from the fovea and encode form and color. Each layer receives signals from only one eye. Layers 2, 3, and 5 receive signals from the temporal hemiretina of the ipsilateral eye, and layers 1, 4, and 6 receive signals from the nasal hemiretina of the contralateral eye.

Neurons in all six layers of the lateral geniculate nucleus project to the primary visual cortex in the occipital lobe via the geniculocalcarine tract (also known as the "optic radiations"). The geniculocalcarine tract can be divided into two main bundles. Geniculate neurons that receive signals from the dorsal hemiretinas (which view the ventral visual field) project to the cortex in the upper (dorsal) bank of the calcarine fissure. Their axons leave the lateral geniculate nucleus in the sublenticular part of the internal capsule and pass lateral to the lateral ventricle before terminating in the visual cortex. Geniculate neurons that receive signals from the ventral hemiretinas (which view the dorsal visual field) project to the cortex in the lower (ventral) bank of the calcarine fissure. Their axons initially course anteriorly in the temporal lobe, then make a U-turn above the inferior horn of the lateral ventricle and swing posteriorly before terminating. The anterior loop of these axons is called "Meyer's loop."

Parvocellular layers in the lateral geniculate nucleus, representing the central retina, project to the posterior aspect of the primary visual cortex, whereas the magnocellular layers, representing the peripheral retina, project to the anterior aspect of the primary visual cortex (see Fig. II–9).

Primary Visual Cortex

The primary visual cortex is mostly buried within the depths of the calcarine fissure. On the medial surface of the hemisphere, the visual cortex extends from the posterior occipital pole forward to the junction of the calcarine fissure with the

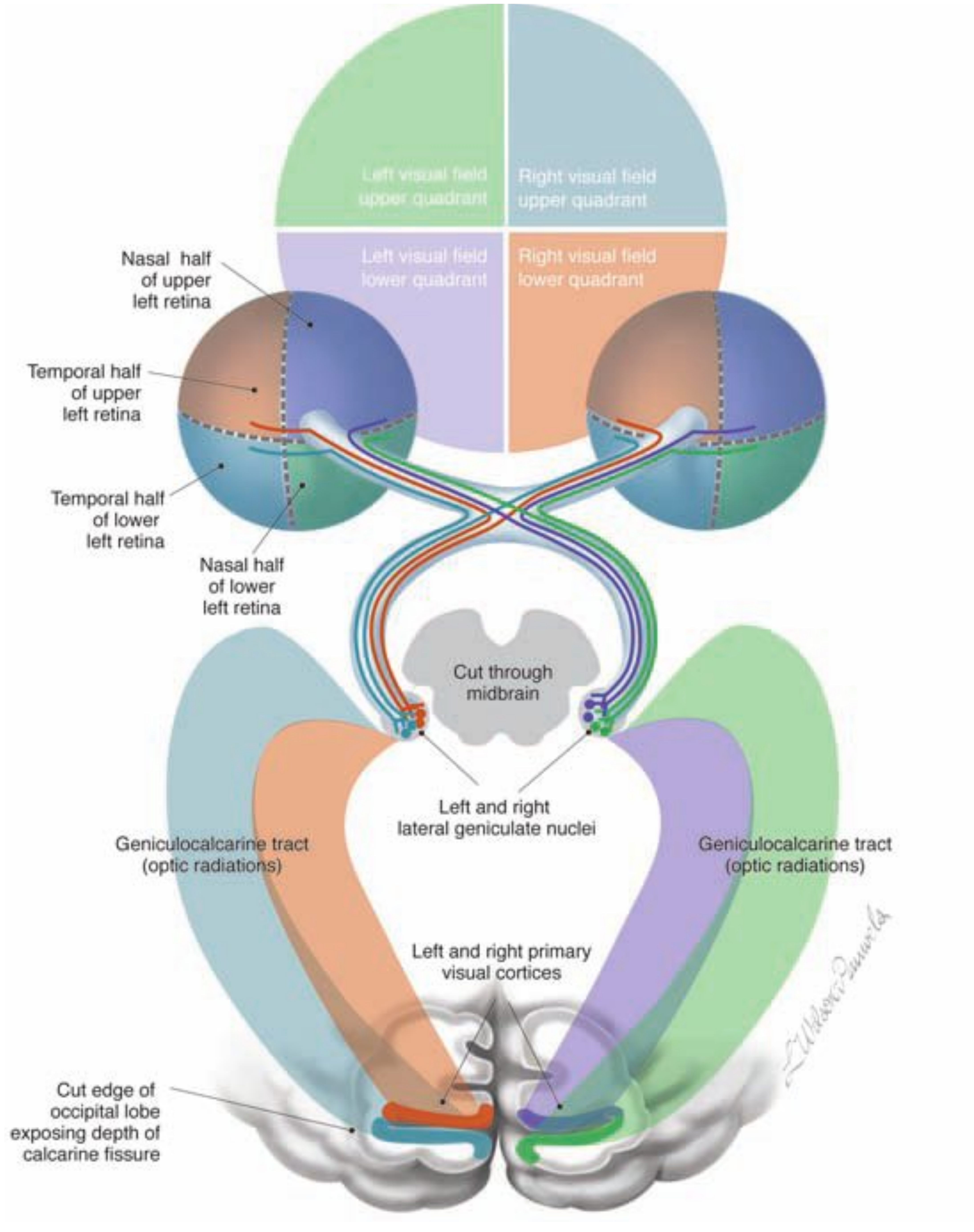

Left visual field upper quadrant
Right visual field upper quadrant
Left visual field lower quadrant
Right visual field lower quadrant
Nasal half of upper left retina
Temporal half of upper left retina
Temporal half of lower left retina
Nasal half of lower left retina
Cut through midbrain
Left and right lateral geniculate nuclei
Geniculocalcarine tract (optic radiations)
Geniculocalcarine tract (optic radiations)
Left and right primary visual cortices
Cut edge of occipital lobe exposing depth of calcarine fissure

FIGURE II–8 The visual pathway. The quadrants of the visual fields are color-coded to show transmission of information to the retinae and to the primary visual cortices where the image is first perceived. (Representation of the foveas are not shown in this illustration, see Figure II–9.)

parieto-occipital fissure. The cortex occupies the upper and lower banks of the calcarine fissure. Each bank receives signals from one-quarter of the retina in both eyes and, therefore, views one-quarter of the visual field. The upper bank of the calcarine fissure forms an image of the contralateral lower visual field from both eyes, and the

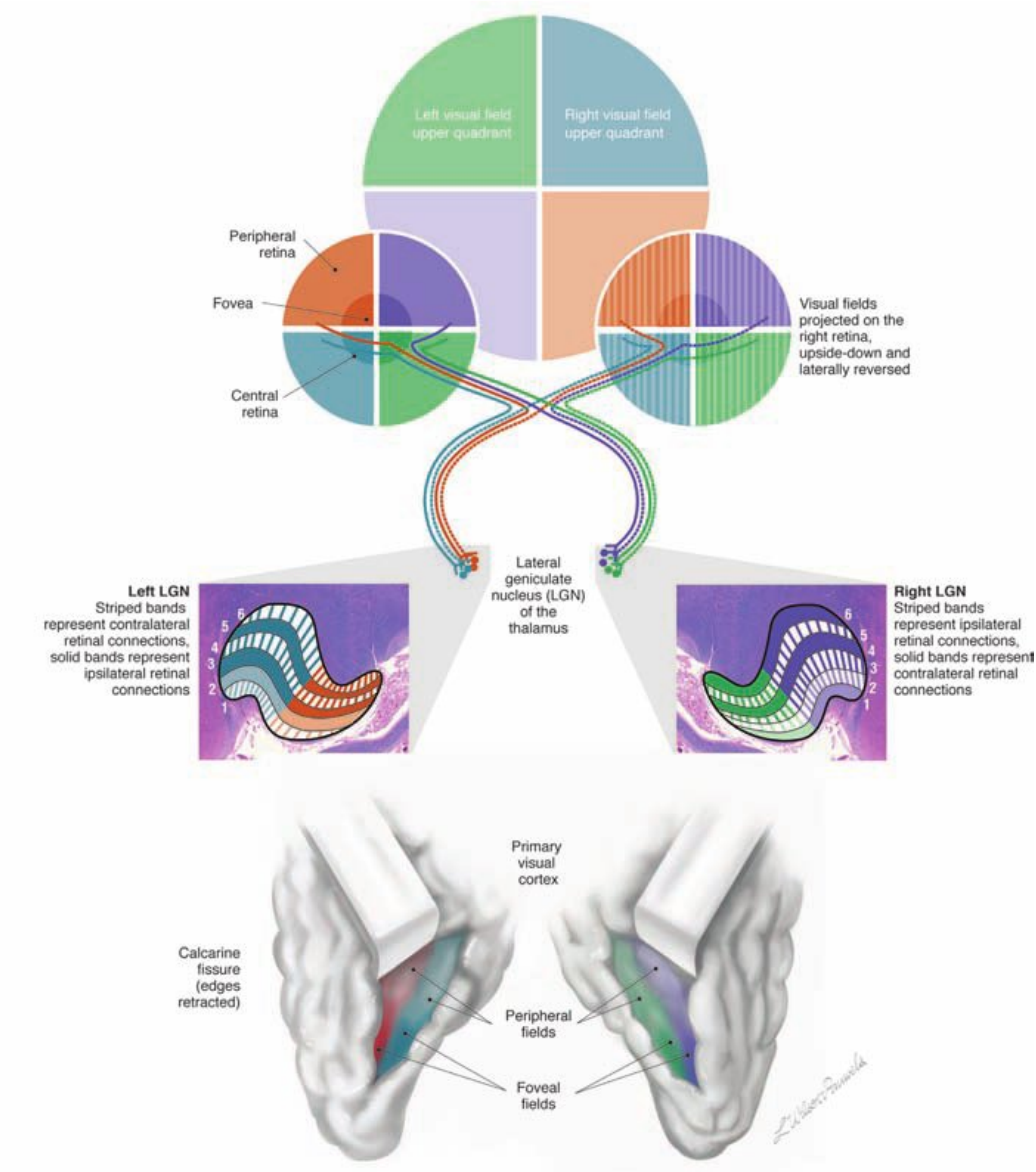

Left visual field upper quadrant
Right visual field upper quadrant
Peripheral retina
Fovea
Central retina
Visual fields projected on the right retina, upside-down and laterally reversed
Lateral geniculate nucleus (LGN) of the thalamus
Left LGN
Striped bands represent contralateral retinal connections, solid bands represent ipsilateral retinal connections
1
2
3
4
5
6
Right LGN
Striped bands represent ipsilateral retinal connections, solid bands represent contralateral retinal connections
6
5
4
3
2
1
Primary visual cortex
Calcarine fissure (edges retracted)
Peripheral fields
Foveal fields

FIGURE II–9 The primary visual cortices surrounding the calcarine fissures receive signals from the four quadrants of the visual fields via the lateral geniculate nuclei. The upper visual fields are mapped below the calcarine fissures and the lower visual fields are mapped above the calcarine fissures.

lower bank of the calcarine fissure forms an image of the contralateral upper visual field in both eyes (see Fig. II–9).

Signals from the primary visual cortex are sent continuously to the visual association cortices for further processing. These signals are used to construct a perceived visual field that is right side up and oriented correctly from left to right. The resolution is perceived to be constant across the field. From the accessory cortices, there are thought to be two principal streams of visual information sent to higher-order cortical areas: a dorsal stream that projects to the parietal lobe for perception of movement and a ventral stream that projects to the ventral temporal lobe for perception of form and color.

CASE HISTORY GUIDING QUESTIONS

1. Why was Meredith's vision blurred and color discrimination reduced in her right eye?
2. What is the pupillary light reflex?
3. Why was there little constriction of Meredith's pupils when a bright light was shone in her right eye?
4. What other lesions of the visual pathway result in visual loss?

1. Why was Meredith's vision blurred?

Only the central retina has high-enough resolution to allow for reading and recognition of faces (Fig. II–10A). This area also has a high concentration of cones, which are responsible for color vision. The inflammation in Meredith's right optic nerve (optic neuritis) interferes with transmission of signals from the retina to the lateral geniculate nucleus. Because a high proportion of the optic nerve axons originate in the fovea, Meredith's central vision is considerably compromised, and vision in her right eye is therefore blurry (see Fig. II–10B). Discrimination of color is also impaired and red appears less intense. This can be assessed quickly by looking for red desaturation. A bright red object such as a bottle cap is presented to each eye and the patient is asked to comment on whether the color red is dimmer or brighter relative to the other eye. The affected eye typically perceives the red as dimmer. This can be more formally assessed using Ishihara plates, but often these are not readily available. Testing of color desaturation is not a routine part of the cranial nerve (CN) examination and is conducted only if the examiner thinks that CN II is impaired.

2. What is the pupillary light reflex?

The pupillary light reflex evolved to control the amount of light entering the eye. In dim light. the pupil is dilated to allow maximal entry of light; however, as the light gets brighter, the pupil gets smaller. The pupillary light reflex involves two cranial

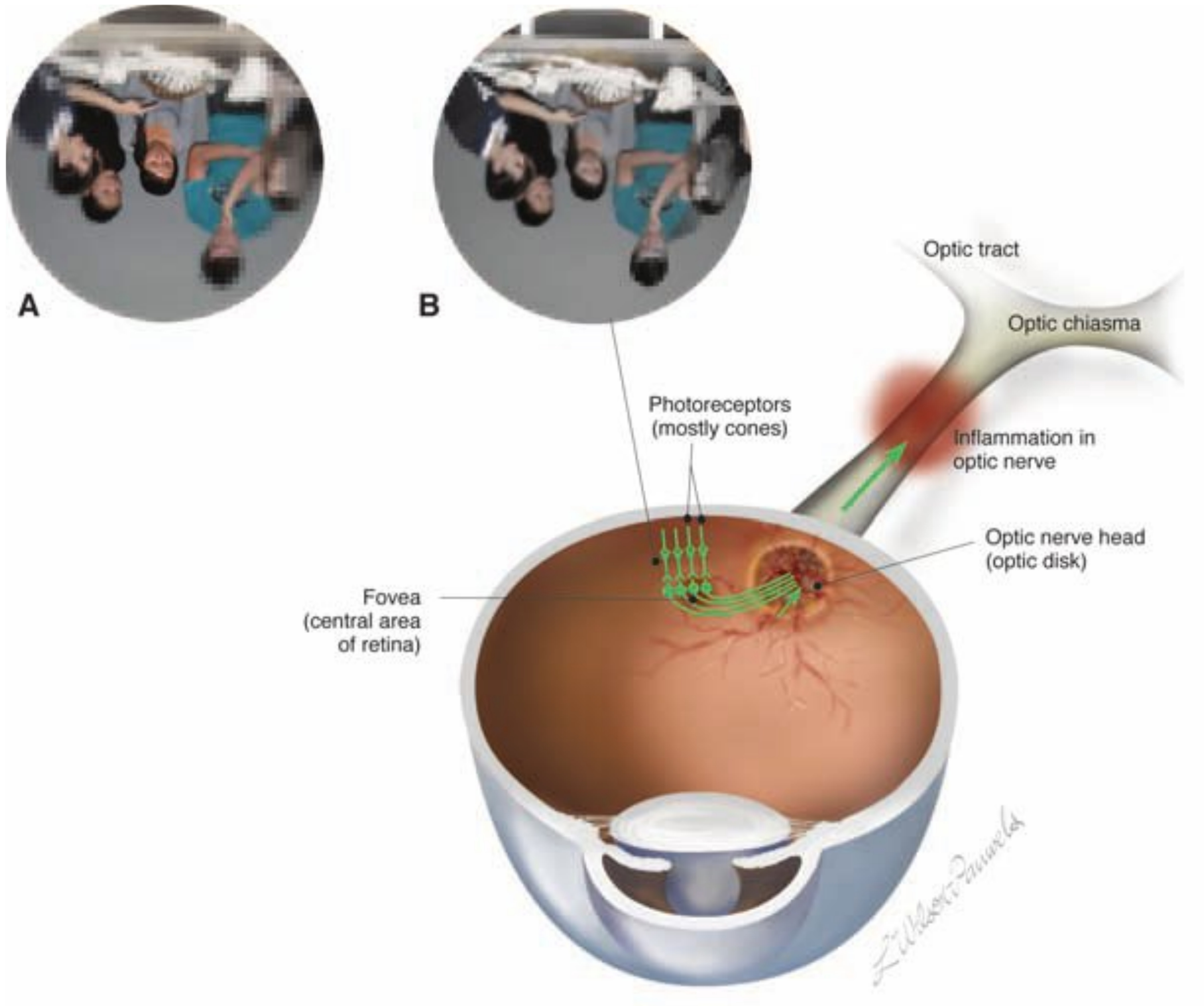


FIGURE II–10 Normally Meredith's central vision has high acuity and color information **(A)**. Inflammation in her right optic nerve interferes with the transmission of signals from the retina to the visual cortex. Since half of the axons in the optic nerve carry signals from the fovea, the major change Meredith perceives is a loss of acuity and color in her central vision **(B)**.

nerves: the optic nerve (CN II) forms the sensory limb by carrying the sensory signal to the brain stem and the oculomotor nerve (CN III) forms the motor limb by carrying motor signals to the pupillary constrictor muscle.

Light entering the eyes causes signals to be sent to both the ipsilateral and the contralateral pretectal olivary nuclei in the pretectal region of the midbrain. Many of these signals originate from the intrinsically photosensitive retinal ganglion cells described in the *Specialist Comment 1*, page 29. The pretectal olivary nucleus cells, in turn, send signals bilaterally to the Edinger-Westphal nuclei. Visceral motor signals arising in the Edinger-Westphal nuclei are sent along preganglionic parasympathetic axons in the oculomotor nerves to the ciliary ganglia. Postganglionic axons leave the ciliary ganglia via 6 to 10 short ciliary nerves to enter the eyes at their pos-

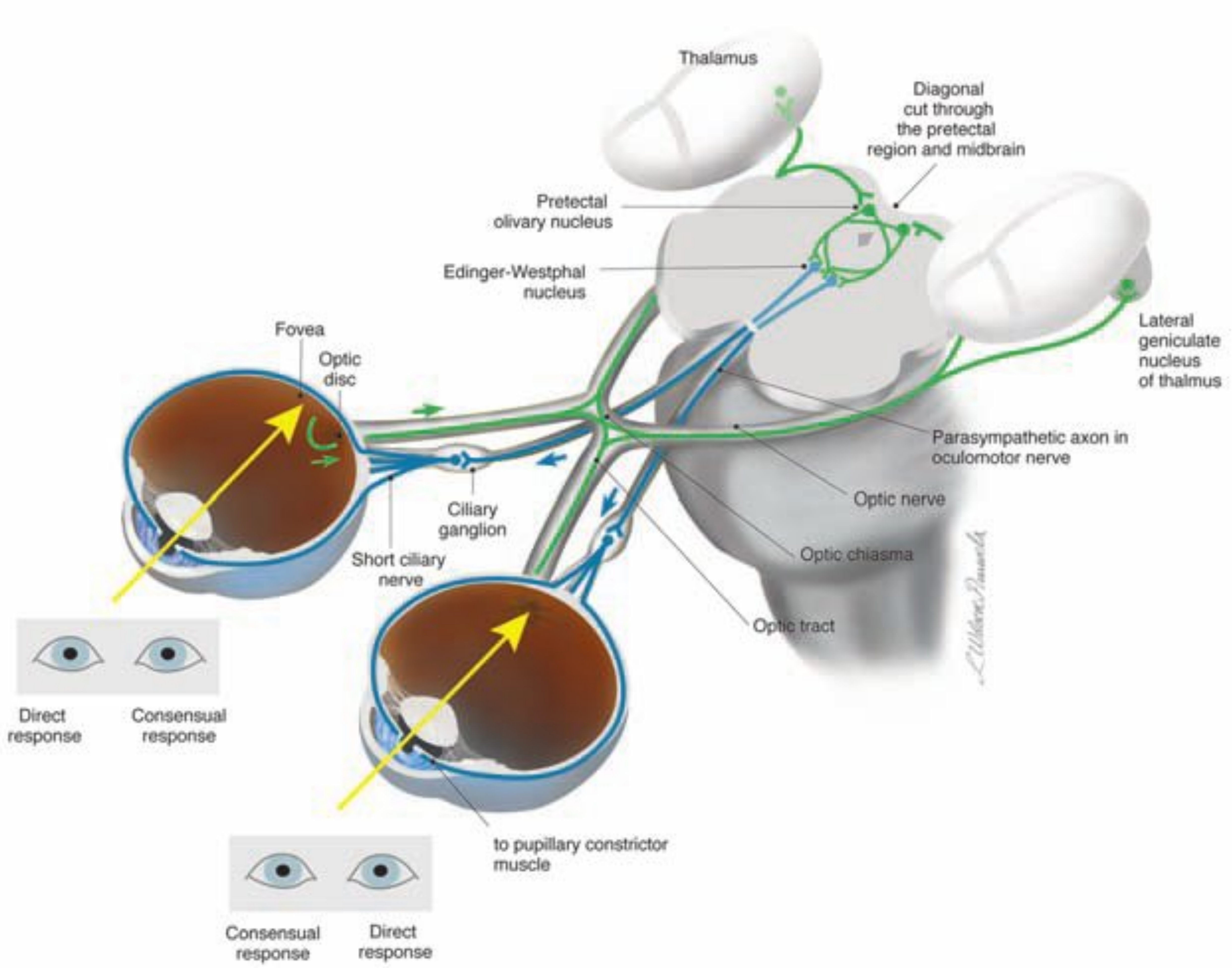

Thalamus
Diagonal cut through the pretectal region and midbrain
Pretectal olivary nucleus
Edinger-Westphal nucleus
Fovea
Optic disc
Lateral geniculate nucleus of thalmus
Parasympathetic axon in oculomotor nerve
Optic nerve
Ciliary ganglion
Short ciliary nerve
Optic chiasma
Optic tract
Direct response
Consensual response
to pupillary constrictor muscle
Consensual response
Direct response

FIGURE II–11 Pupillary light reflex. Light shone in either eye elicits pupillary constriction in the same eye (direct response) and in the opposite eye (consensual response) via the pathway shown.

terior aspects near the origin of the optic nerves. Within the globe, the nerves run forward between the choroid and the sclera to terminate in the constrictor pupillae muscles of the iris (Fig. II–11).

In the normal reflex, light shone in either eye causes constriction of the pupil in the same eye (the direct light reflex) and also in the other eye (the consensual light reflex).

3. Why was there little constriction of Meredith's pupils when a bright light was shone in her right eye?

When a light was shone into Meredith's right eye, transmission of the signal to her midbrain was reduced or blocked because of inflammation of the optic nerve (Fig. II–12). The defect in the sensory limb of the reflex pathway resulted in poor direct and consensual pupillary responses *relative to* the response produced by light shone in the intact eye. This is referred to as a *relative* afferent pupillary defect, also referred to as a "Marcus Gunn pupil." Compare Meredith's afferent pupillary defect with Werner's motor pupillary defect in Chapter III, Figure III–11.

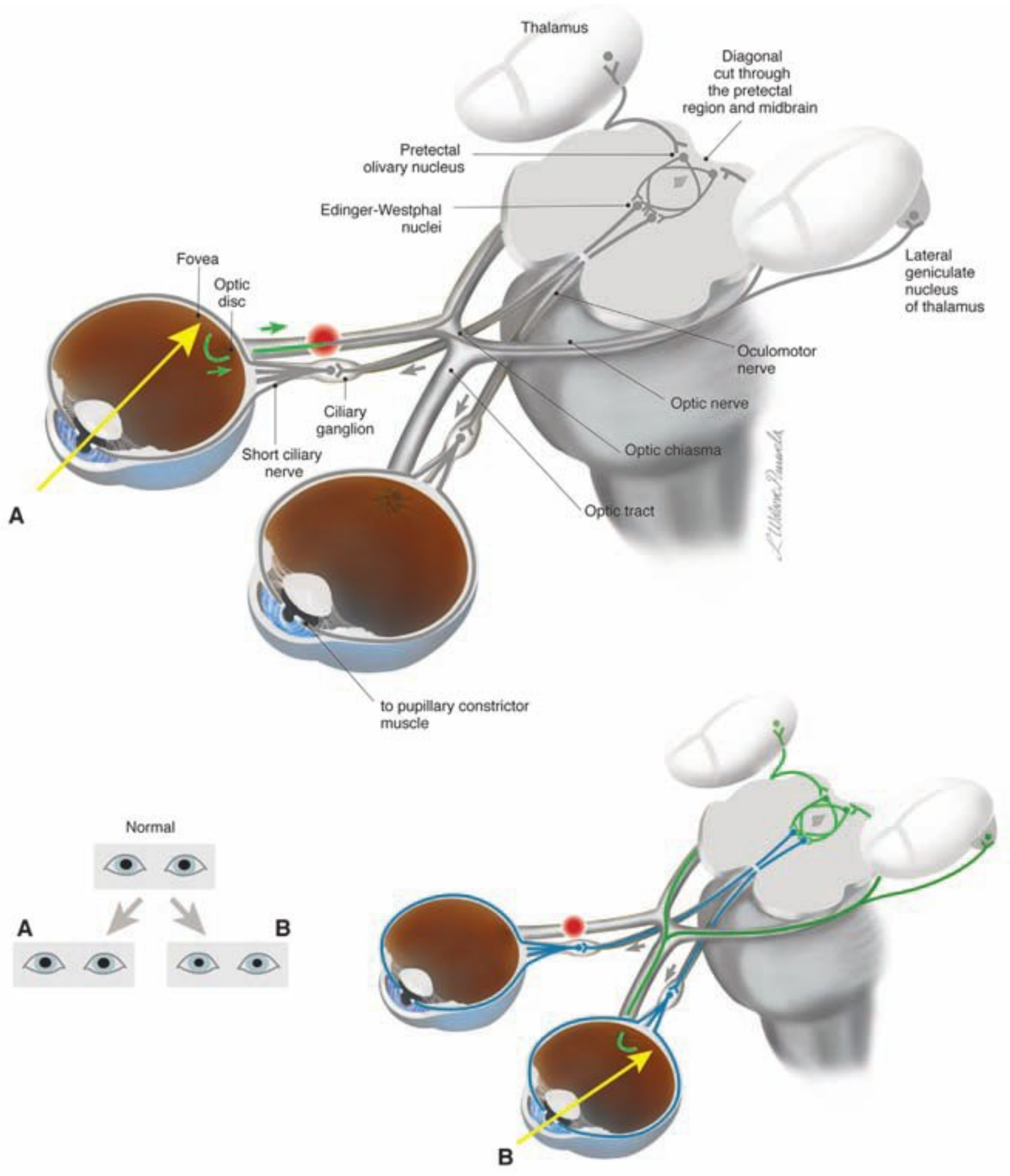

Thalamus
Diagonal cut through the pretectal region and midbrain
Pretectal olivary nucleus
Edinger-Westphal nuclei
Fovea
Optic disc
Lateral geniculate nucleus of thalamus
Oculomotor nerve
Optic nerve
Ciliary ganglion
Short ciliary nerve
Optic chiasma
Optic tract
A
to pupillary constrictor muscle
Normal
A
B
B

FIGURE II–12 Relative afferent pupillary defect.

A. Light shown in Meredith's right eye results in the absence of direct (right) and consensual (left) pupillary constriction.

B. Light shown in Meredith's left eye results in pupillary constriction in the left eye (direct response) and pupillary constriction in the opposite eye (consensual response).

4. What other lesions of the visual pathway result in visual loss?

Lesions in different sites in the optic pathway produce different patterns of visual loss. Typical sites for damage and the visual loss that each produces are shown in Figures II–13 to II–16.

Clinically, it is useful to group visual pathway lesions into three categories:

- *Anterior to the Chiasma*

  Damage to the light-transmitting parts of the eye, the retina, or the optic nerve results in visual loss in the affected eye only, which is described as a "monocular visual loss" (see Fig. II–13).

Specialist Comment 3

Until recently, it was thought that a proportion of the axons that cross in the chiasma swing forward into the opposite optic nerve for a short distance before turning posteriorly and entering the optic tract. This group of axons, known as "Wilbrand's knee," was thought to explain visual loss in the opposite eye when the optic nerve was damaged close to the chiasma. Wilbrand performed his studies on patients who had had one eye removed so that he could identify definitively the axons in the stump of the optic nerve as arising from the contralateral eye. It is now known that the "knee" is an artifact of monocular enucleation and is not found in the normal optic chiasma (Horton, 1997).

- *At the Chiasma*

  Damage to the optic chiasma usually results in loss of vision from both eyes, depending on which axons are affected. Figure II–14 illustrates damage to the midline of the chiasma, which causes a loss of the peripheral fields in both eyes. This is called "bitemporal hemianopia" (see Fig. II–14).

- *Posterior to the Chiasma*

  Damage to the optic tracts, lateral geniculate body, optic radiations, or visual cortices results in visual loss from both eyes within the contralateral visual field. This type of visual field loss is known as "homonymous hemianopia" (see Fig. II–15). "Macular sparing" refers to sparing of central vision within the hemianopic (blind) hemifield. Macular sparing signifies sparing of the posterior pole of the occipital lobe, which receives macular (central vision) information from the contralateral hemisphere. Remember that the macula is the area of the central retina that includes the fovea.

- *Lesions of Meyer's Loop*

  Damage to Meyer's loop results in homonymous quadrantanopia, or loss of the contralateral upper visual field in both eyes (see Fig. II–16)

- *Partial Loss of Axons along the Visual Pathway*

  Lesions that involve only a subgroup of axons in the visual pathway produce scotomas (i.e., partial loss of the visual field). Scotomas are also called "blind spots." Meredith has a scotoma in her right eye.

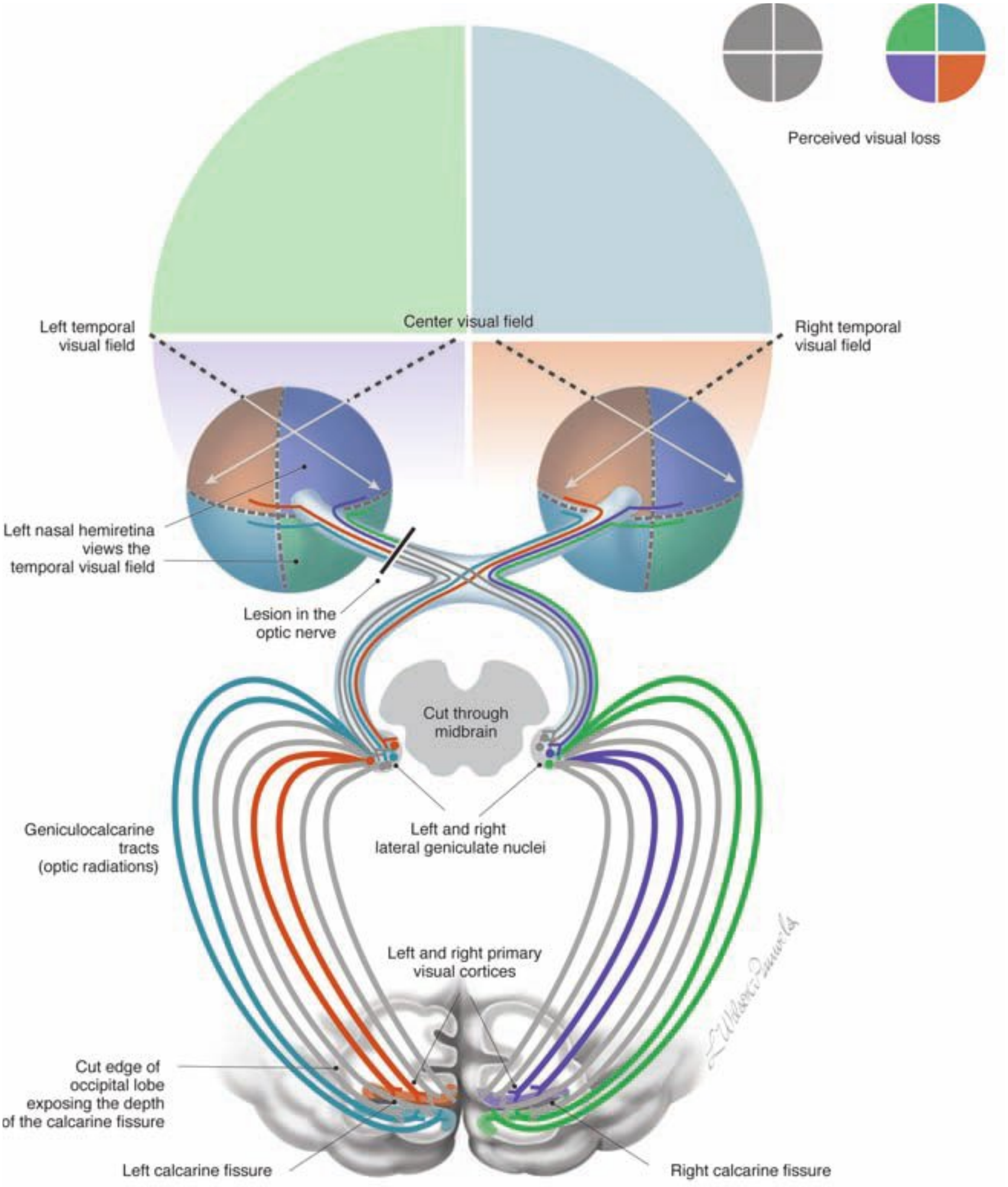

Perceived visual loss
Left temporal visual field
Center visual field
Right temporal visual field
Left nasal hemiretina views the temporal visual field
Lesion in the optic nerve
Cut through midbrain
Geniculocalcarine tracts (optic radiations)
Left and right lateral geniculate nuclei
Left and right primary visual cortices
Cut edge of occipital lobe exposing the depth of the calcarine fissure
Left calcarine fissure
Right calcarine fissure

FIGURE II–13 Monocular visual loss
Damage to the visual pathway anterior to the chiasm results in visual loss in the affected eye only. Damage to the left optic nerve, as illustrated, results in a loss of vision in the left eye.

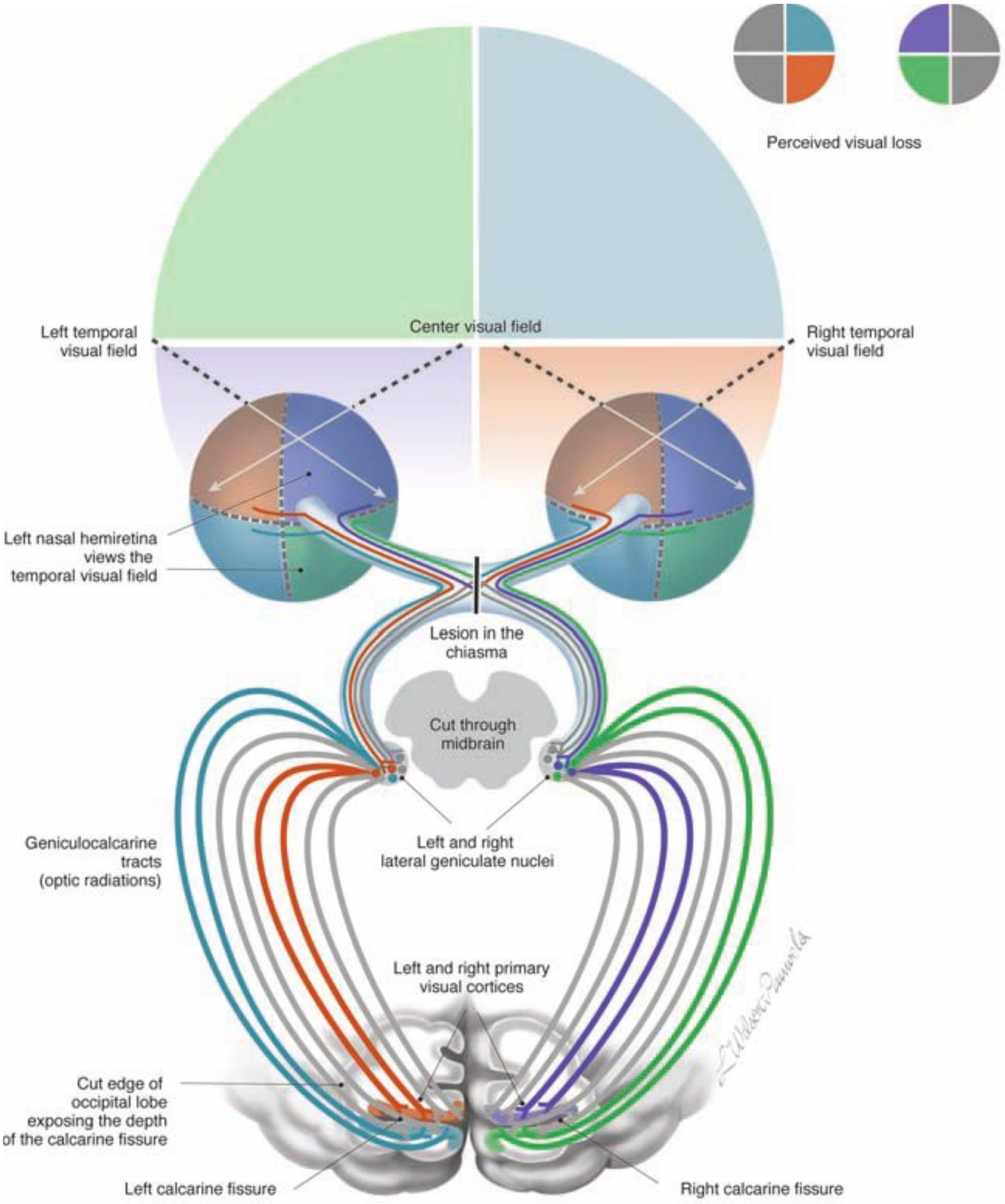

Perceived visual loss
Left temporal visual field
Center visual field
Right temporal visual field
Left nasal hemiretina views the temporal visual field
Lesion in the chiasma
Cut through midbrain
Left and right lateral geniculate nuclei
Geniculocalcarine tracts (optic radiations)
Left and right primary visual cortices
Cut edge of occipital lobe exposing the depth of the calcarine fissure
Left calcarine fissure
Right calcarine fissure

FIGURE II–14 Bitemporal hemianopia
Lesions to the visual pathway at the chiasma results in visual loss in both eyes. Damage to the midline of the chiasma, as illustrated, interferes with transmission of signals from the nasal hemiretinae, which view the temporal visual fields in both eyes.

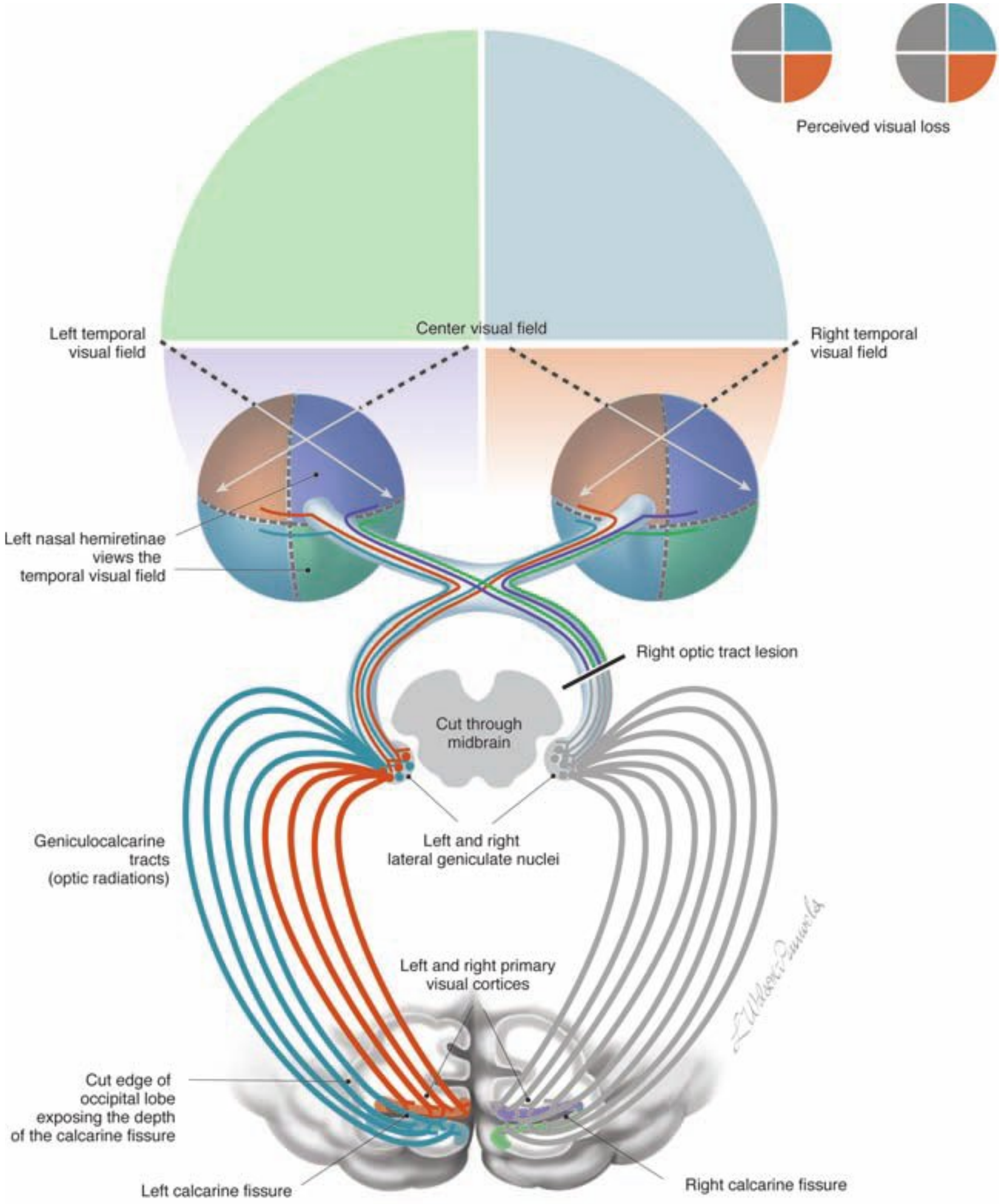

Perceived visual loss
Left temporal visual field
Center visual field
Right temporal visual field
Left nasal hemiretinae views the temporal visual field
Right optic tract lesion
Cut through midbrain
Geniculocalcarine tracts (optic radiations)
Left and right lateral geniculate nuclei
Left and right primary visual cortices
Cut edge of occipital lobe exposing the depth of the calcarine fissure
Left calcarine fissure
Right calcarine fissure

FIGURE II–15 Homonymous hemianopia.
Lesions in the visual pathway posterior to the chiasma result in loss of signals from the contralateral fields in both eyes. For example, a lesion lo the right optic tract, as illustrated, eliminates perception of the left visual field.

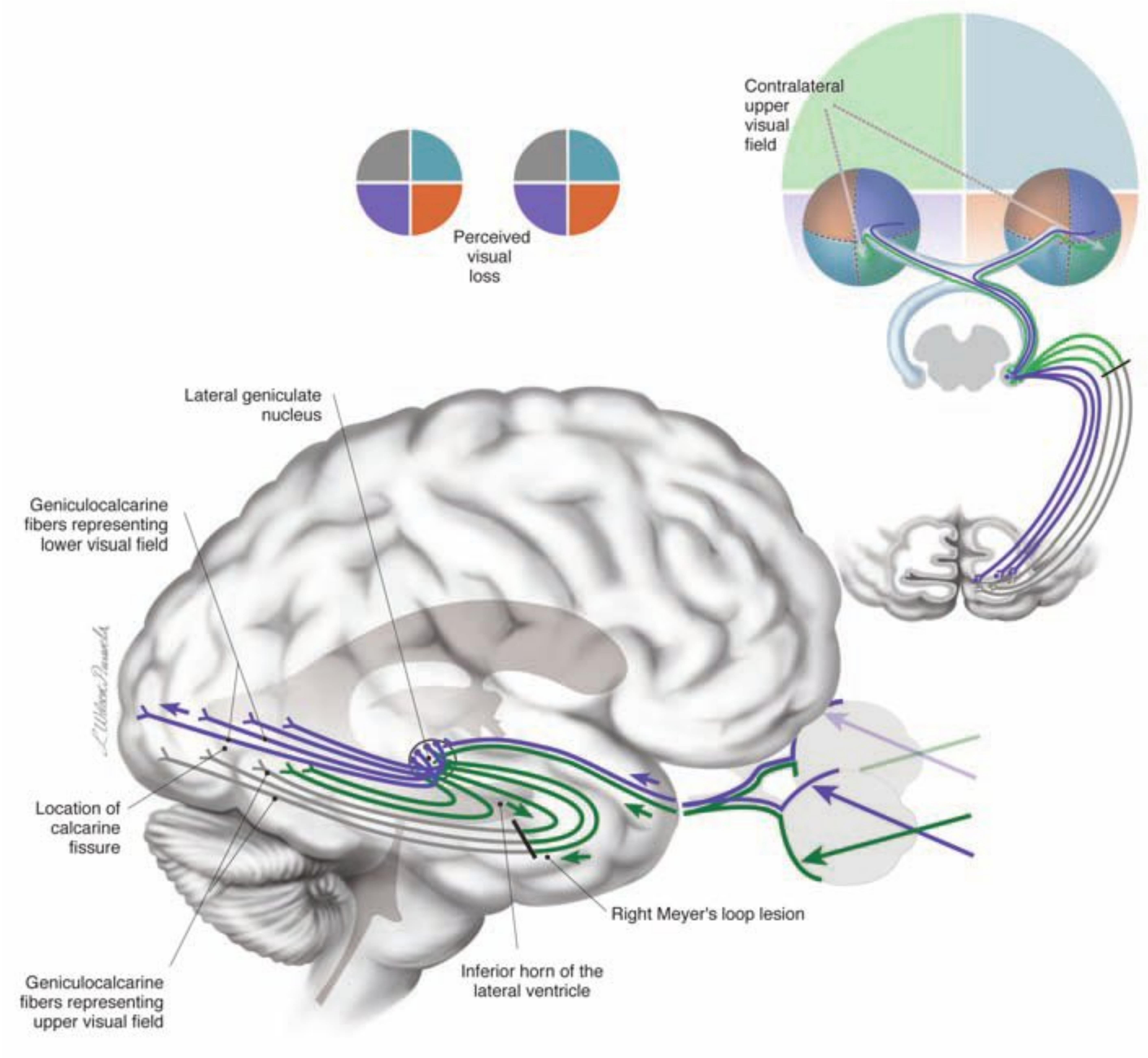

Contralateral upper visual field
Perceived visual loss
Lateral geniculate nucleus
Geniculocalcarine fibers representing lower visual field
Location of calcarine fissure
Right Meyer's loop lesion
Inferior horn of the lateral ventricle
Geniculocalcarine fibers representing upper visual field

FIGURE II–16 Left Upper Quadrantanopia
Lesions to the visual pathway in the right Meyer's loop results in the loss of the contralateral upper quadrant of the visual field in both eyes.

CLINICAL TESTING

Examination of the optic nerve involves four procedures:

- Measurement of visual acuity.
- Testing of visual fields.
- Testing of the pupillary light reflex.
- Visualization of the fundus.

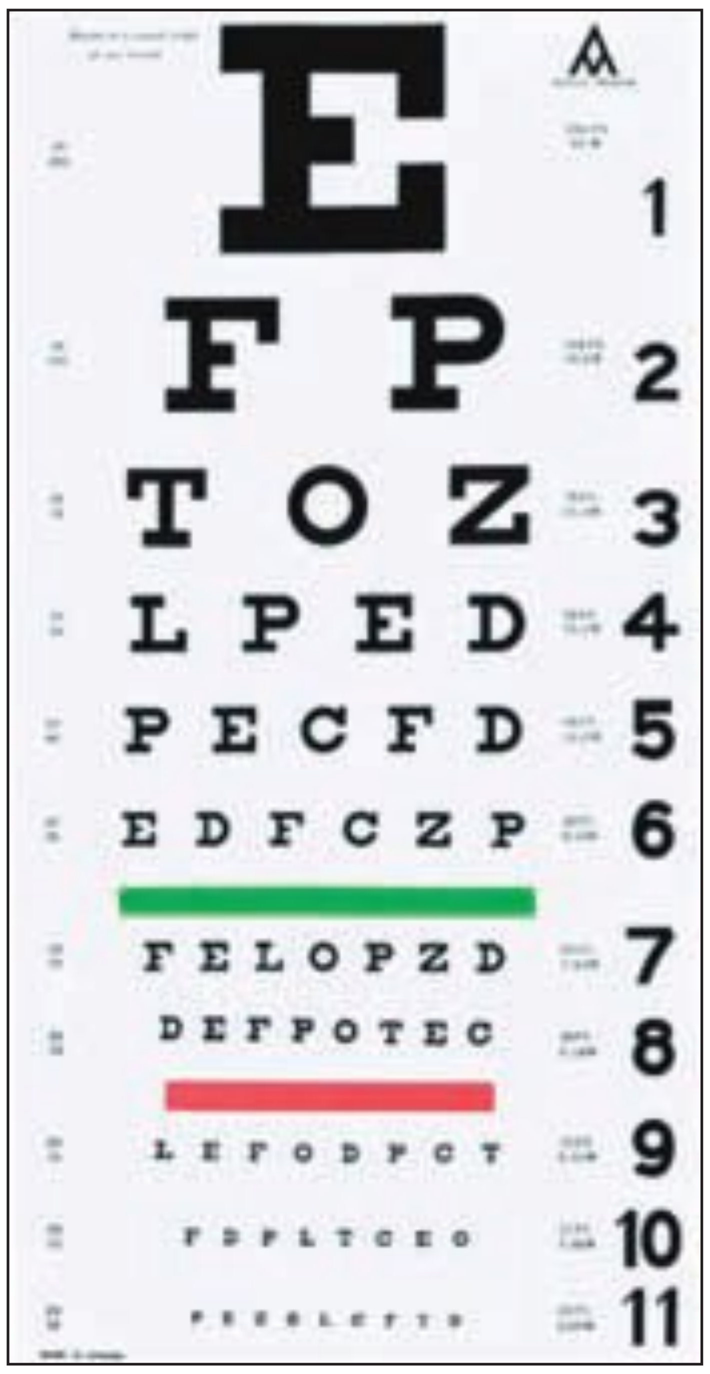
(See also "Cranial Nerves Examination" on website.)

Measurement of Visual Acuity

Visual acuity is best assessed using Snellen's chart[‡] (Fig. II–17) at 20 feet; however, a hand-held visual acuity card can be used at the bedside. The patient may wear his or her glasses, and each eye is tested separately. Visual acuity is a test of macular function. The macula gives rise to the majority of the optic nerve fibers; therefore, inflammation or a lesion of the optic nerve can result in significant loss of visual acuity.

Testing of Visual Fields

Visual field testing (Fig. II–18) at the bedside involves comparison of the examiner's visual fields with those of the patient. Sit directly across from the patient. Instruct



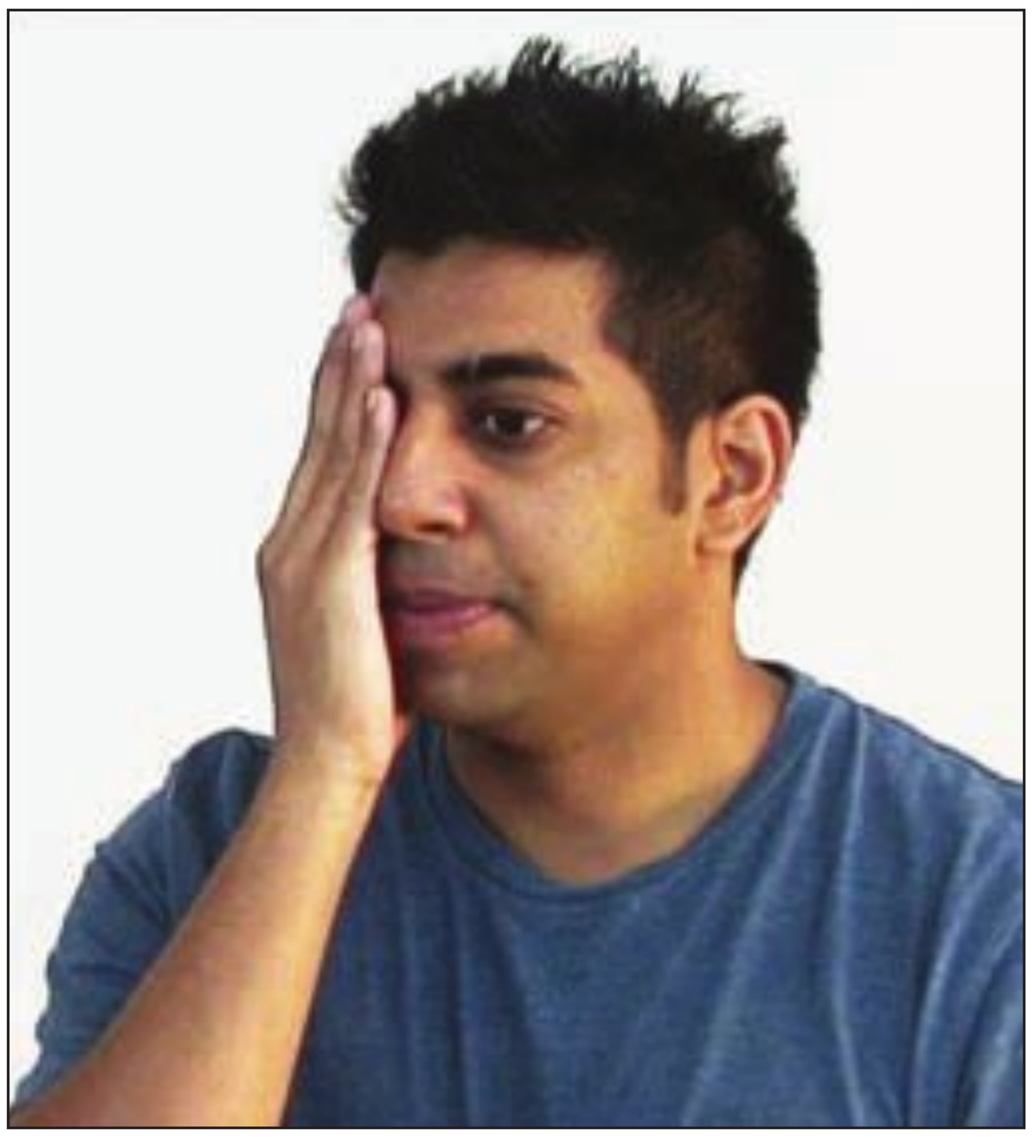


FIGURE II–17 Acuity is tested using Snellen's chart.

[‡] Named for the Dutch ophthalmologist Herman Snellen, who developed the chart in 1862.

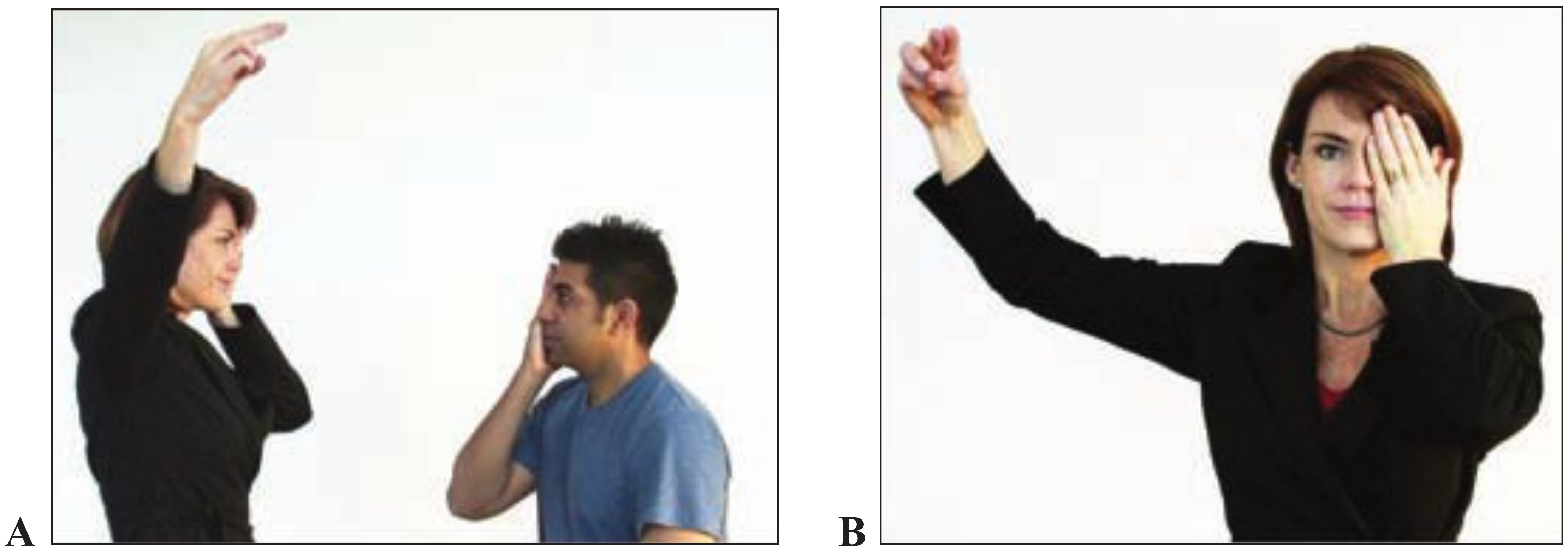


FIGURE II–18 Testing of the visual field. **A**, Testing the upper left quadrant of the patient's left eye (patient's right eye and physician's left eye are covered). **B**, Patient's view of the physician testing the upper left quadrant of the patient's left eye.

the patient to focus on your nose and then ask the patient to cover his or her right eye (see Fig. II–18). You, in turn, cover your left eye and, with your arm fully extended, bring your index finger from beyond the periphery of vision toward the center of the visual field (see Fig. II–18B). The patient is asked to indicate when the finger is first seen. Both you and the patient should see the finger at the same time. Your finger should be brought in obliquely in all four quadrants. The procedure is repeated for the other eye. A field defect is likely present if the patient fails to see your finger once it is in your own visual field.

A full understanding of the anatomy of the visual pathway is necessary to interpret the results of visual field testing (see Figs. II–13 and II–16).

Testing of the Pupillary Light Reflex

The pupillary light reflex relies on the integrity of both CN II (the sensory pathway) and the parasympathetic nerve fibers that travel with CN III (the motor pathway). A beam of light is shone directly on one pupil. If both the sensory and the motor pathways are intact, the ipsilateral pupil constricts; this is the direct response. The contralateral pupil should also constrict. This is the indirect (consensual) response (see Fig. II–11). If the patient has a sensory pupillary defect, light shone in the affected eye will cause only mild constriction of both pupils; however, when the light is shone in the normal pupil, there will be a strong constriction of both pupils. As the light is swung back and forth from eye to eye, the pupils will alternately constrict, as light is shone in the good eye, and dilate from the constricted state, as light is shone in the bad eye. This is referred to as the "swinging flashlight" test.

Visualization of the Fundus

Visualization of the fundus involves the use of an ophthalmoscope (Fig. II–19A). The examination is performed in a dimly lit room so that the patient's pupils are maximally dilated. Ask the patient to focus on an object in the distance. This helps

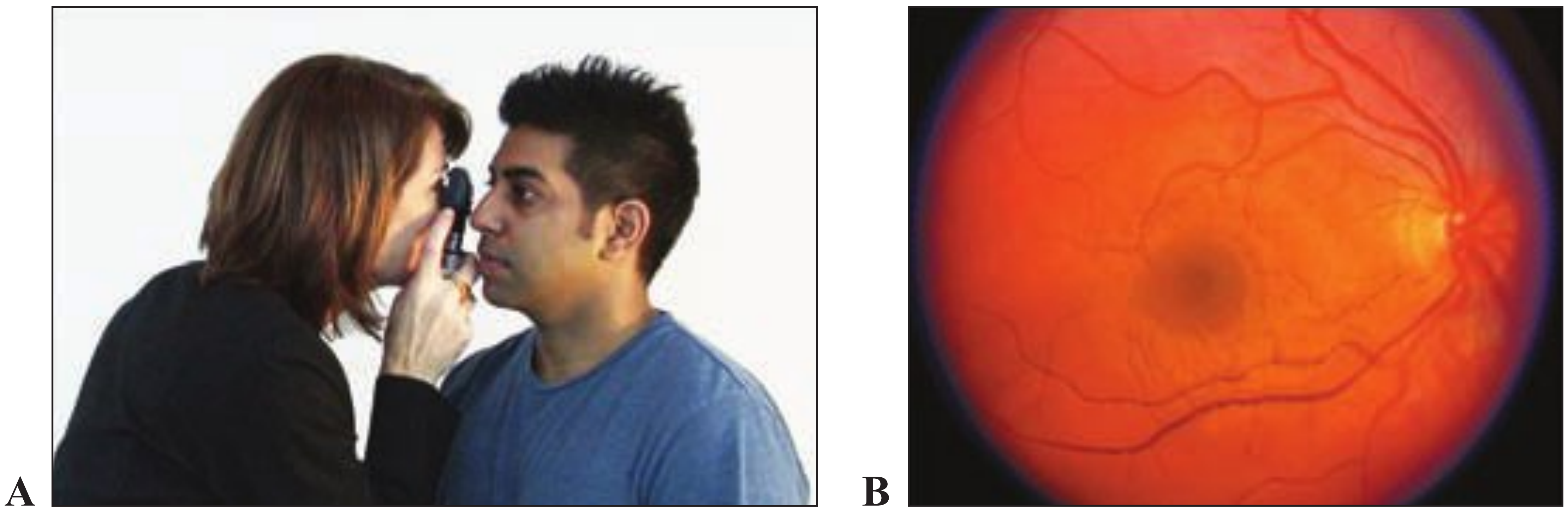


FIGURE II–19 Examination of the fundus. **A**, The physician is examining the patient's right fundus using an ophthalmoscope. **B**, Normal fundus of the right eye. (Fundus photograph courtesy of Dr. R. Buncic.)

to keep the eyes still and allows for better visualization of the fundus. The first thing to look at is the disc (optic nerve head). The margins of the disc should be sharp. Blurring of the disc margins is seen with raised intracranial pressure, and disc pallor is an indication of optic atrophy. You should also note the optic cup. The optic cup is a depression in the center of the disk from which vessels emerge. Take a close look at the vessels; venous pulsations can be seen in 85% to 90% of patients. They disappear if the intracranial pressure is raised. Lastly, look at the retina and macula for further evidence of disease. Ophthalmoscopy takes a lot of practice and we recommend that you practice on your classmates to become familiar with the appearance of a normal fundus (see Fig. II–19B).

ADDITIONAL RESOURCES

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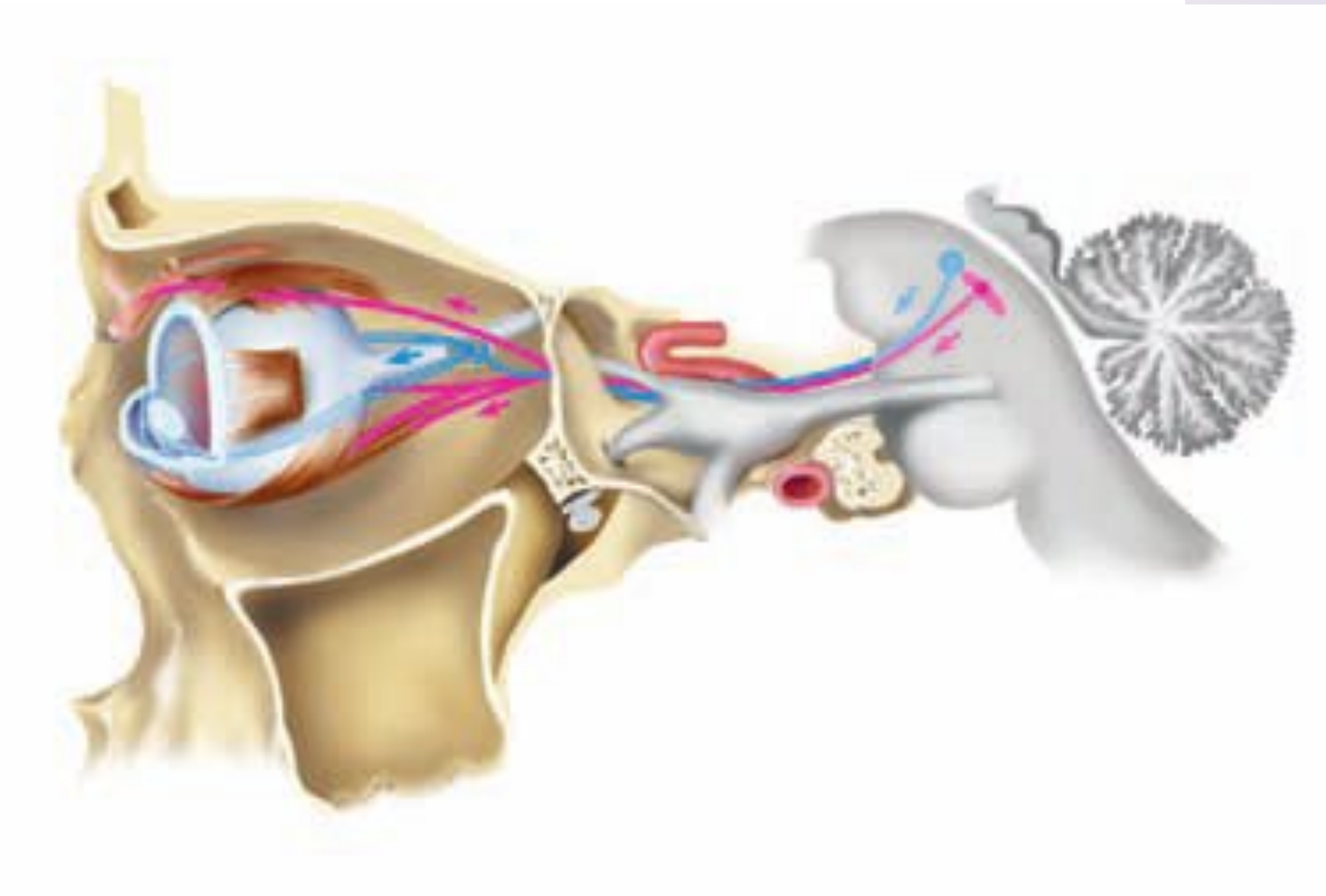

III

Oculomotor Nerve

CASE HISTORY

Werner is a 54-year-old gentleman whose favorite pastime is working in his garden. One afternoon, he experienced a sudden headache while lifting a heavy potted plant. This headache was the worst he had ever experienced and he started to vomit. Because the headache was sudden and very severe, Werner went to the emergency department of the nearest hospital.

In the emergency department, Werner was drowsy and had a stiff neck, although he was rousable and able to answer questions and follow commands. When a bright light was shone into his left eye, his left pupil constricted briskly, but the right pupil remained dilated. When the light was shone directly into his right eye, his right pupil remained dilated and his left pupil constricted. Werner also had a droopy right eyelid, and when he was asked to look straight ahead, his right eye deviated slightly down and to the right. Werner complained of double vision and recalled that he had experienced some sensitivity to light in his right eye during the 2 weeks preceding this event. Close examination of Werner's eye movements revealed that he could move his left eye in all directions, but had difficulty with movements of his right eye. With his right eye, Werner was able to look to the right (abduct) but he could not look to the left (adduct). He was not able to look directly up or down. Werner's other cranial nerves (CNs) were tested and found to be functioning normally.

The emergency physician was concerned that Werner might have experienced a subarachnoid hemorrhage. A computed tomography (CT) scan of his head was performed, and this demonstrated blood in the subarachnoid space. A cerebral angiogram was done, which demonstrated an aneurysm (a localized widening of an artery due to weakness in the vessel wall) of the right posterior communicating artery. Werner subsequently underwent neurosurgery to have the aneurysm repaired.

ANATOMY OF THE OCULOMOTOR NERVE

As its name implies, the oculomotor nerve plays a major role in eye movement (Table III–1). Its somatic motor component innervates four of the six extraocular (extrinsic) muscles, and its visceral motor component innervates the intrinsic ocular muscles (constrictor pupillae and the ciliary muscle). The nerve also innervates the levator palpebrae superioris muscle, which elevates the upper eyelid (Fig. III–1).

TABLE III–1 Components, Nuclei, Ganglia, and Function of the Oculomotor Nerve (CN III)

Component	Nucleus	Ganglion	Function
Somatic motor (efferent)	Oculomotor nucleus		To innervate: levator palpebrae superioris, superior rectus, medial rectus, inferior rectus and inferior oblique muscles
Parasympathetic motor (visceral efferent)	Edinger-Westphal nucleus	Ciliary ganglion	To innervate constrictor pupillae and ciliary muscles

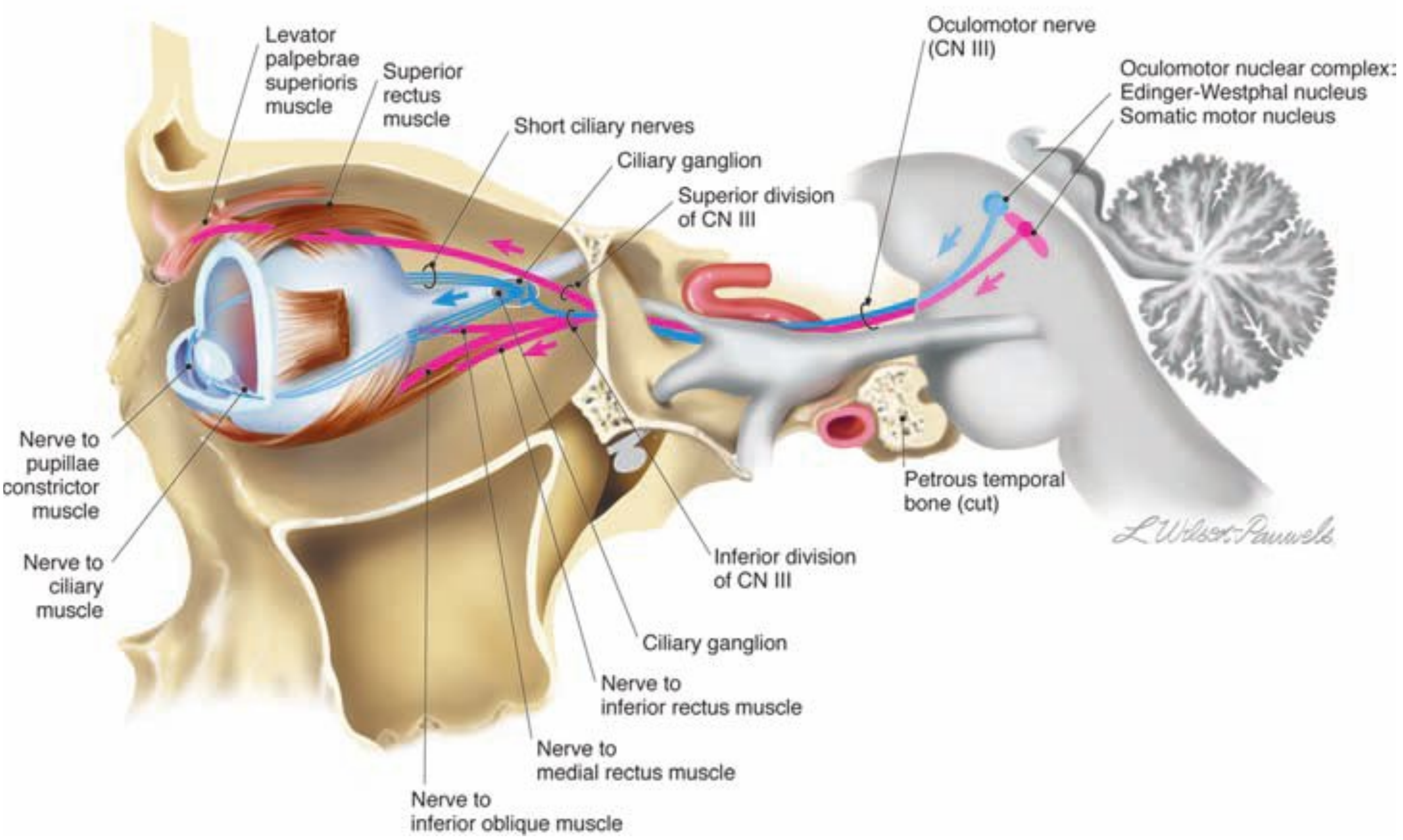
Levator palpebrae superioris muscle
Superior rectus muscle
Short ciliary nerves
Ciliary ganglion
Superior division of CN III
Oculomotor nerve (CN III)
Oculomotor nuclear complex:
Edinger-Westphal nucleus
Somatic motor nucleus
Nerve to pupillae constrictor muscle
Nerve to ciliary muscle
Petrous temporal bone (cut)
L Wilson-Pauwels
Inferior division of CN III
Ciliary ganglion
Nerve to inferior rectus muscle
Nerve to medial rectus muscle
Nerve to inferior oblique muscle

FIGURE III–1 Overview of the oculomotor nerve.

The oculomotor complex consists of the somatic motor subnuclei and the Edinger-Westphal nucleus, which is composed of visceral motor neurons. The complex is located in the midbrain at the level of the superior colliculus near the midline and just ventral to the cerebral aqueduct. It is bounded laterally and inferiorly by the medial longitudinal fasciculus. Axons of both nuclei course ventrally in the midbrain to form the oculomotor nerve, which emerges from the interpeduncular fossa on the ventral aspect of the midbrain (Fig. III–2). After passing between the posterior cerebral and the superior cerebellar arteries (Fig. III–3), the nerve courses anteriorly.

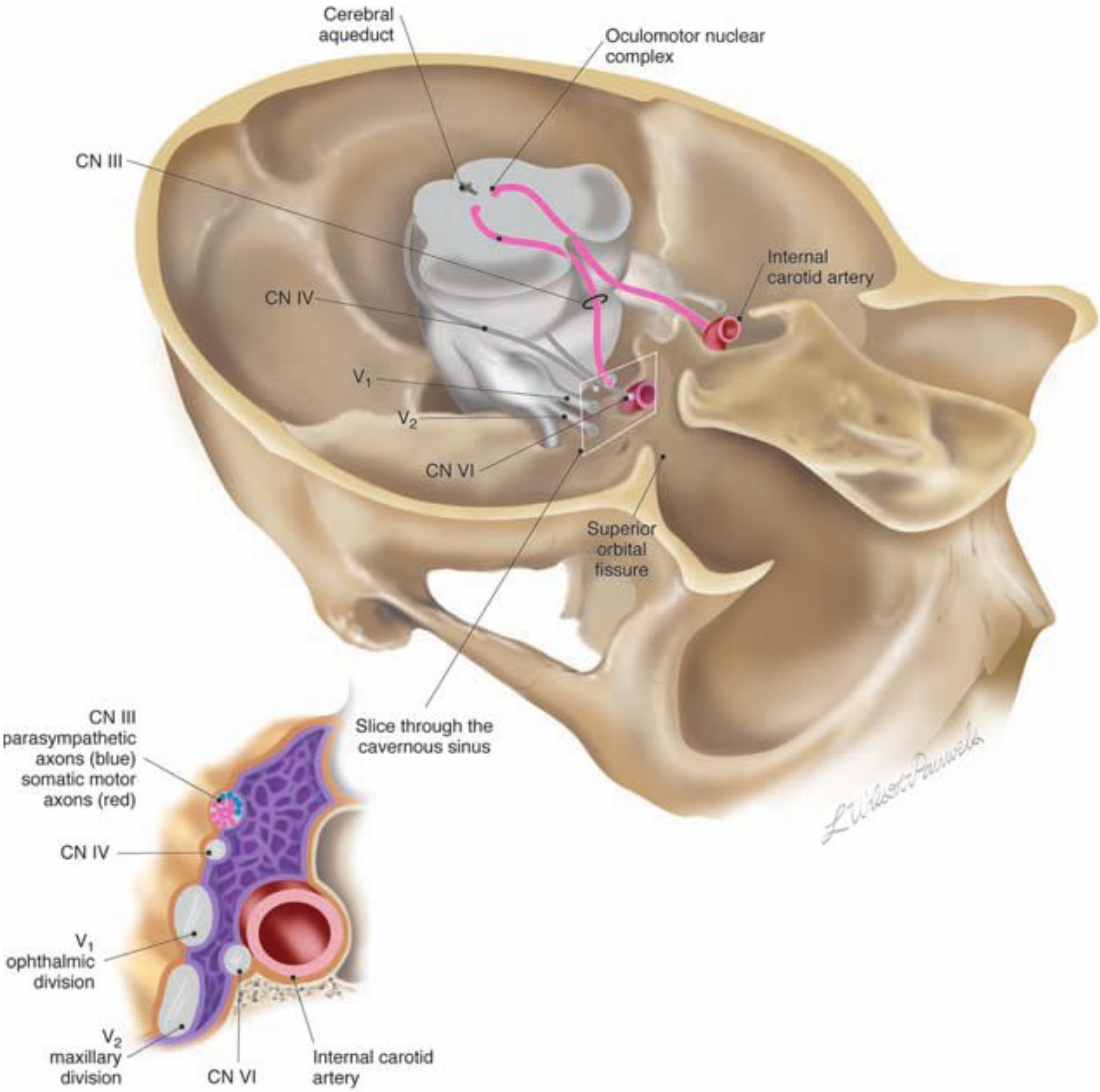

FIGURE III–2 Oculomotor nerve. The inset illustrates the anatomic relationship of CN III to other structures as it courses through the cavernous sinus. Note in cranial nerve III, the parasympathetic axons are clustered superio-medially on the surface of the nerve.

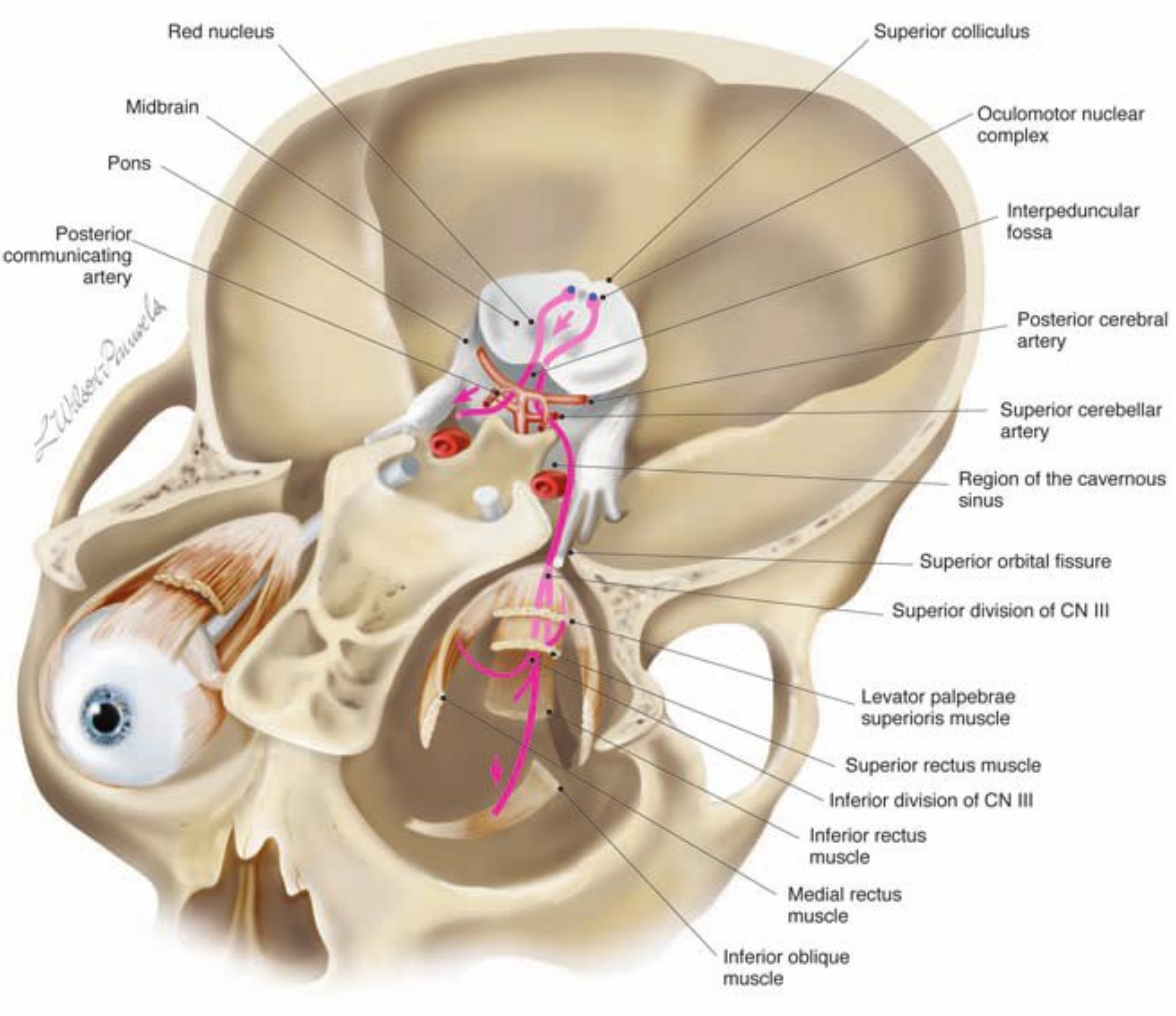
Red nucleus
Midbrain
Pons
Posterior communicating artery
L Wilson-Pauwels
Superior colliculus
Oculomotor nuclear complex
Interpeduncular fossa
Posterior cerebral artery
Superior cerebellar artery
Region of the cavernous sinus
Superior orbital fissure
Superior division of CN III
Levator palpebrae superioris muscle
Superior rectus muscle
Inferior division of CN III
Inferior rectus muscle
Medial rectus muscle
Inferior oblique muscle

FIGURE III–3 The somatic motor component of the oculomotor nerve.

It pierces the dura and enters the cavernous sinus where it runs along the lateral wall of the sinus (see Fig. III–2 inset) just superior to the trochlear nerve (CN IV) and then continues forward toward the superior orbital fissure. The nerve splits into a smaller superior division and a larger inferior division and then passes through the superior orbital fissure within the tendinous ring (annulus of Zinn) (Fig. III–3).

The superior division innervates the superior rectus and the levator palpebrae superioris muscles. The inferior division innervates the medial rectus, inferior rectus, and inferior oblique muscles. The visceral motor axons run with the nerve to the inferior oblique muscle for a short distance, then leave it to terminate in the ciliary ganglion. Postganglionic axons leave the ciliary ganglion as 8 to 10 short ciliary nerves to enter the eye at the posterior aspect near the exit of the optic nerve (see Fig. III–1).

Somatic Motor (Efferent) Component

The oculomotor nucleus is located in the tegmentum of the midbrain at the level of the superior colliculus. It is generally accepted that subnuclei within the oculomotor nucleus supply individual muscles. The lateral part of the oculomotor complex is formed by the lateral subnuclei, which from dorsal to ventral, supply the ipsilateral inferior rectus, inferior oblique, and medial rectus muscles. The medial subnucleus supplies the contralateral superior rectus muscle, and the central subnucleus (a midline mass of cells at the caudal end of the complex) supplies the levators palpebrae superioris bilaterally (Fig. III–4).

Somatic motor neuron axons leave the oculomotor nuclear complex and course ventrally in the tegmentum of the midbrain through the medial portion of the red nucleus and the medial aspect of the cerebral peduncle to emerge in the interpeduncular fossa at the junction between the midbrain and the pons. They pass through the lateral wall of the cavernous sinus and superior orbital fissure to enter the orbit (see Fig. III–3).

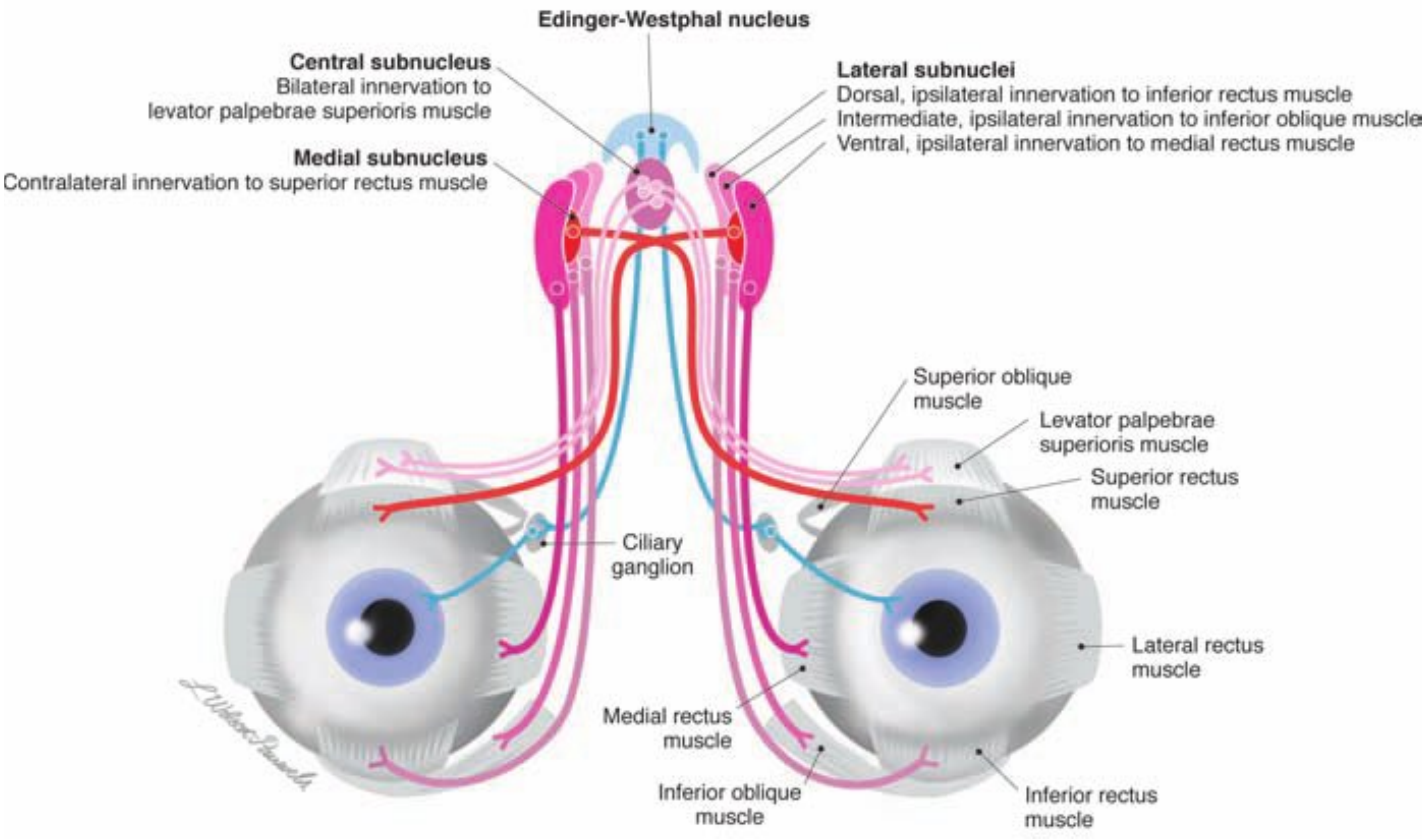
Edinger-Westphal nucleus
Central subnucleus
Bilateral innervation to levator palpebrae superioris muscle
Medial subnucleus
Contralateral innervation to superior rectus muscle
Lateral subnuclei
Dorsal, ipsilateral innervation to inferior rectus muscle
Intermediate, ipsilateral innervation to inferior oblique muscle
Ventral, ipsilateral innervation to medial rectus muscle
Superior oblique muscle
Levator palpebrae superioris muscle
Superior rectus muscle
Ciliary ganglion
Lateral rectus muscle
Medial rectus muscle
Inferior oblique muscle
Inferior rectus muscle

FIGURE III–4 Oculomotor nuclear complex and schematic innervation of extraocular muscles (the functions of the Edinger-Westphal nucleus are discussed with the parasympathetic motor component of cranial nerve III).

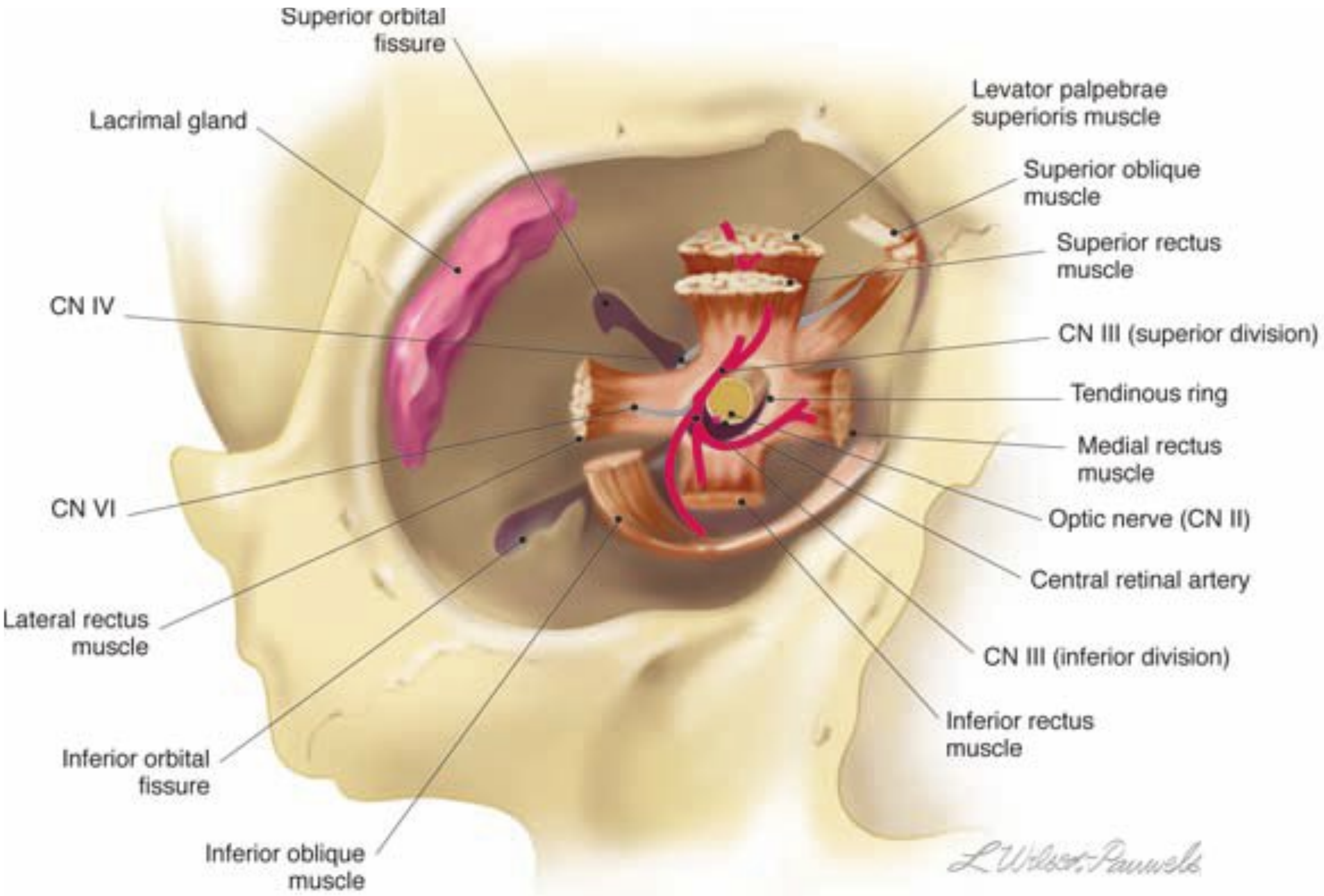

FIGURE III–5 Apex of the right orbit, illustrating the tendinous ring and the somatic motor component of cranial nerve III.

As the somatic motor axons enter the orbital cavity, they branch into superior and inferior divisions (Fig. III–5). The superior division ascends lateral to the optic nerve to supply the superior rectus and levator palpebrae superioris muscles. The inferior division divides into three branches that supply the inferior rectus, inferior oblique, and medial rectus muscles. The muscles are innervated on their ocular surfaces, except for the inferior oblique, whose branch enters the posterior border of the muscle (see Fig. III–5).

The primary actions of the extraocular muscles innervated by CN III (Fig. III–6) are

- Medial rectus muscle: adduction.
- Superior rectus muscle: elevation.
- Inferior rectus muscle: depression.
- Inferior oblique muscle: excyclotorsion.

The combination of these muscles plus the superior oblique (CN IV) and lateral rectus (CN VI) muscles enable eye movements around the three axes of movement shown in Figure III–7. For a more detailed description of the actions of the extraocular muscles, see Chapter XIII.

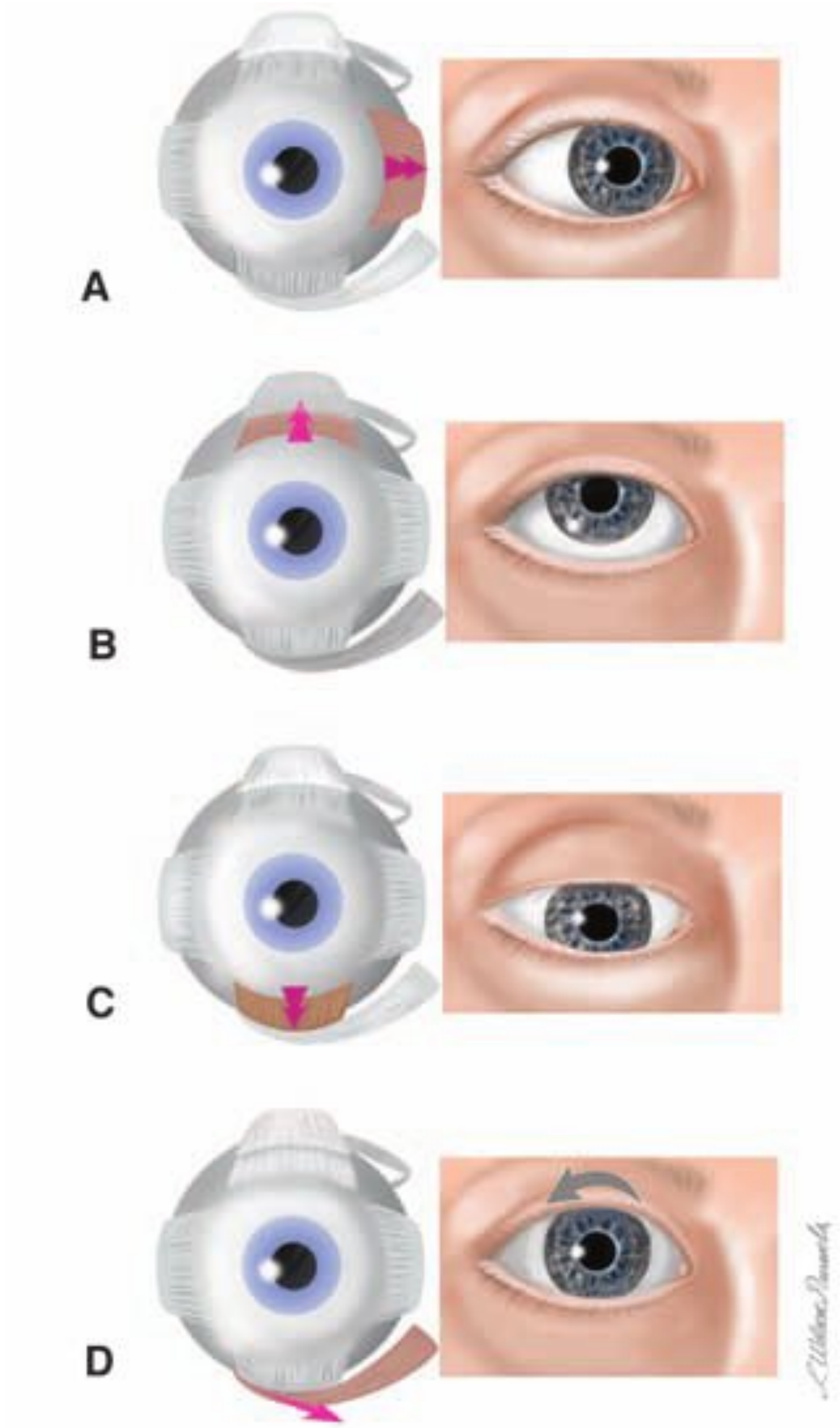

FIGURE III–6 Primary actions of the extraocular muscles innervated by cranial nerve III: **A**, Adduction–medial rectus muscle. **B**, Elevation–superior rectus muscle. **C**, Depression–inferior rectus muscle. **D**, Excyclotorsion–inferior oblique muscle.

Eyelids

The eyelids move with the eye in both upward and downward gaze. The upper eyelid (palpebra superioris) is elevated in upward gaze by its own muscle, the levator palpebra superioris. The levator originates from the orbital roof and the tendinous ring just superior to the origin of the superior rectus muscle (Fig. III–8). Its course through the orbit approximately parallels that of the superior rectus muscle, and it inserts via a broad aponeurosis (tendon) into the skin and tarsus of the upper eyelid. Both the levator and the superior rectus muscles are innervated by the superior division of the oculomotor nerve. Furthermore, the connective tissues of the levator and

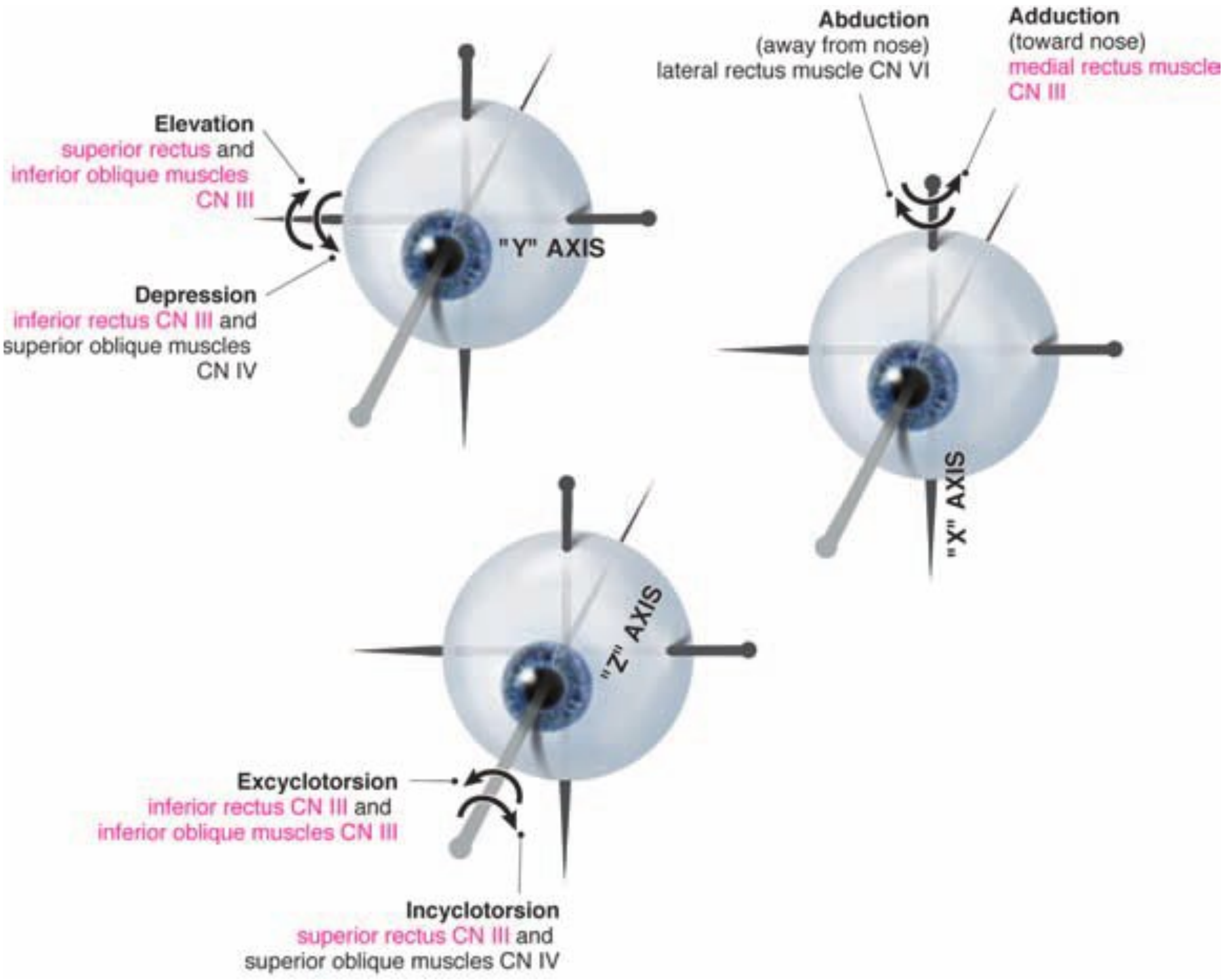

FIGURE III–7 Right eye movements around the “X,” “Y,” and “Z” axes (movements driven by cranial nerve III are highlighted in pink).

the superior rectus muscle are fused. Their movements, therefore, are closely coordinated such that when the eye is elevated, the upper eyelid moves upward as well.

The lower eyelid (palpebra inferioris) does not have its own muscle to effect downward movement of the lower lid in downward gaze. Instead, an extension of the tendon of the inferior rectus muscle reaches forward to insert on the inferior edge of the tarsus of the lower lid. When the inferior rectus muscle is activated to lower the eye, it pulls the lower eye lid downward as well, so that the lid remains clear of the line of sight.

Both eyelids are also moved by bands of smooth muscle, which originate from the tendon of the levator palpebrae superioris muscle in the upper lid, and from the tendinous extension of the inferior rectus muscle in the lower lid. They insert on the superior margin of the upper and inferior margin of the lower tarsal plates. They are called “Müller’s muscles” or the superior and inferior tarsal muscles. The smooth muscle is innervated by sympathetic nerves. For example, activation of the sympathetic nerves, in the flight-or-fight reaction, widens the palpebral fissure and accounts for the “wide-eyed stare” that is characteristic of agitated states.

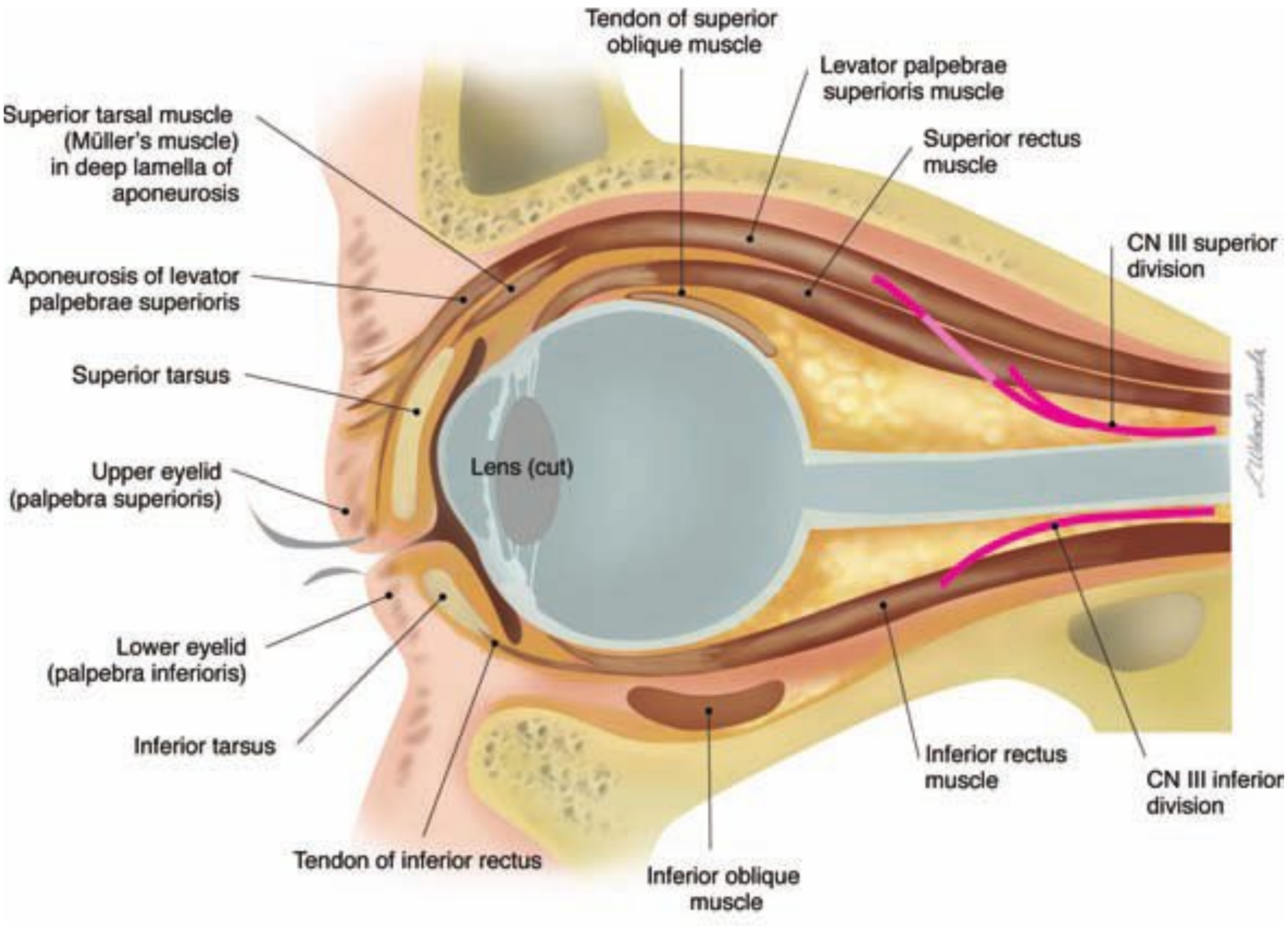
Tendon of superior oblique muscle
Levator palpebrae superioris muscle
Superior tarsal muscle (Müller's muscle) in deep lamella of aponeurosis
Superior rectus muscle
CN III superior division
Aponeurosis of levator palpebrae superioris
Superior tarsus
Upper eyelid (palpebra superioris)
Lens (cut)
Lower eyelid (palpebra inferioris)
Inferior tarsus
Inferior rectus muscle
CN III inferior division
Tendon of inferior rectus
Inferior oblique muscle

FIGURE III–8 Sagittal view of the eye muscles innervated by the occulomotor nerve (CN III) that effect the upper and lower eyelids.

PARASYMPATHETIC MOTOR (VISCERAL EFFERENT) COMPONENT

The Edinger-Westphal (visceral motor) nucleus is located in the midbrain, dorsal to the anterior portion of the oculomotor nucleus (Fig. III–9). Preganglionic visceral motor axons leave the nucleus and course ventrally through the midbrain with the somatic motor axons. The parasympathetic and somatic axons together constitute CN III. The parasympathetic axons are located on the superomedial surface of the nerve (see Fig. III–9). Therefore, when the nerve is compressed on its superomedial aspect, the parasympathetic axons are the first to lose their function. The parasympathetic axons branch from the nerve to the inferior oblique muscle and terminate in the ciliary ganglion near the apex of the orbit (see Fig. III–9).

Postganglionic axons leave the ciliary ganglion as 6 to 10 short ciliary nerves to enter the eye at its posterior aspect near the origin of the optic nerve. Within the globe, the nerves run forward, between the choroid and the sclera, to terminate in the constrictor pupillae muscle and the ciliary muscle (see Fig. III–9). The visceral motor fibers control the tone of their target muscles; therefore, they control the size of the pupil and the shape of the lens.

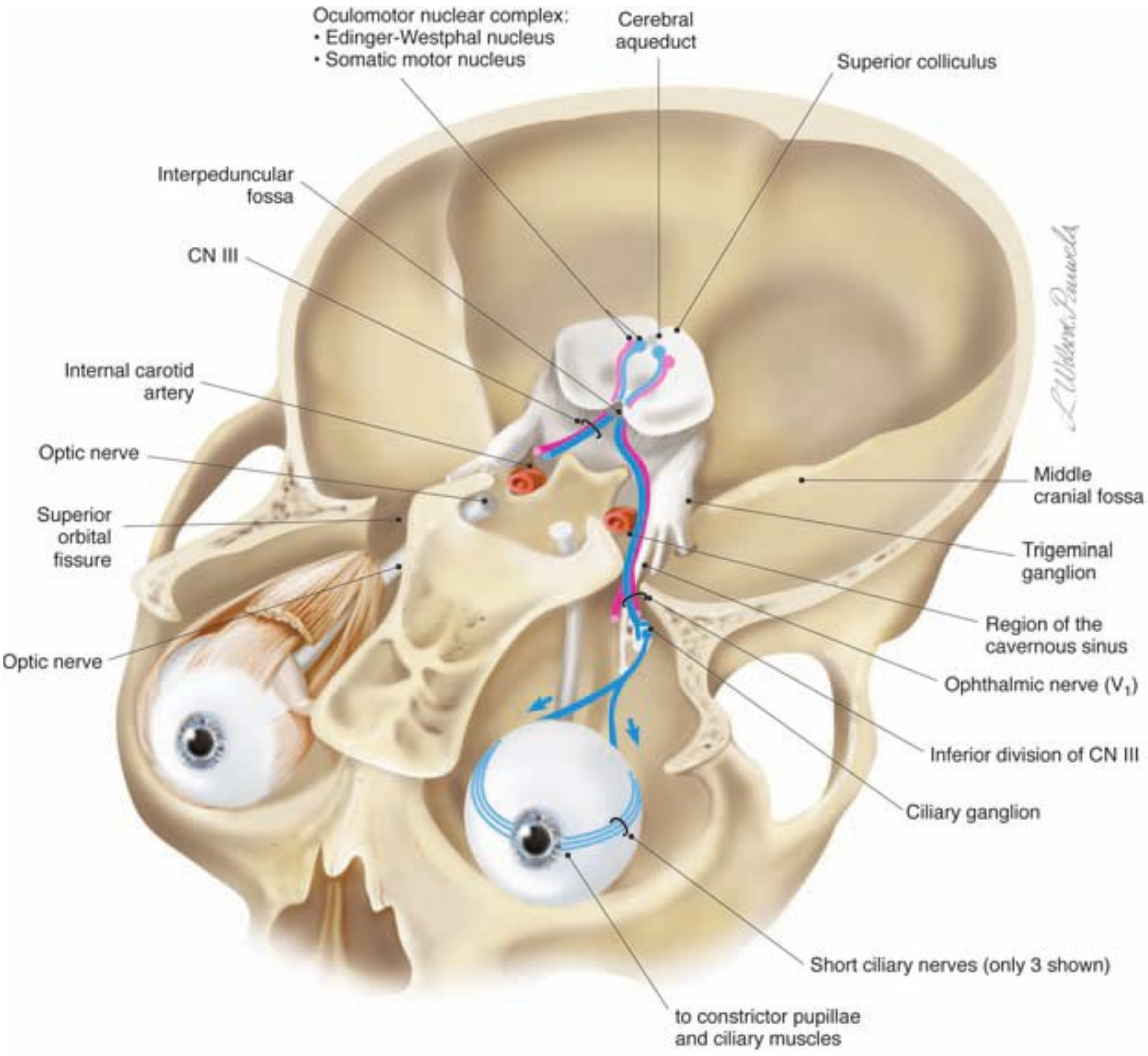

FIGURE III–9 The parasympathetic (visceral motor) component of the oculomotor nerve travels with the somatic motor axons that form the inferior division of cranial nerve III.

Pupillary Light Reflex

The pupillary light reflex is described in Chapter II (see Fig. II–11). The parasympathetic axons that form part of the oculomotor nerve are the motor, limb of this reflex.

Accommodation Reflex

Accommodation is an adaptation of the visual apparatus of the eye for near vision (Fig. III–10). It is accomplished by the following three adjustments in the eye that are often referred to as the “near triad”:

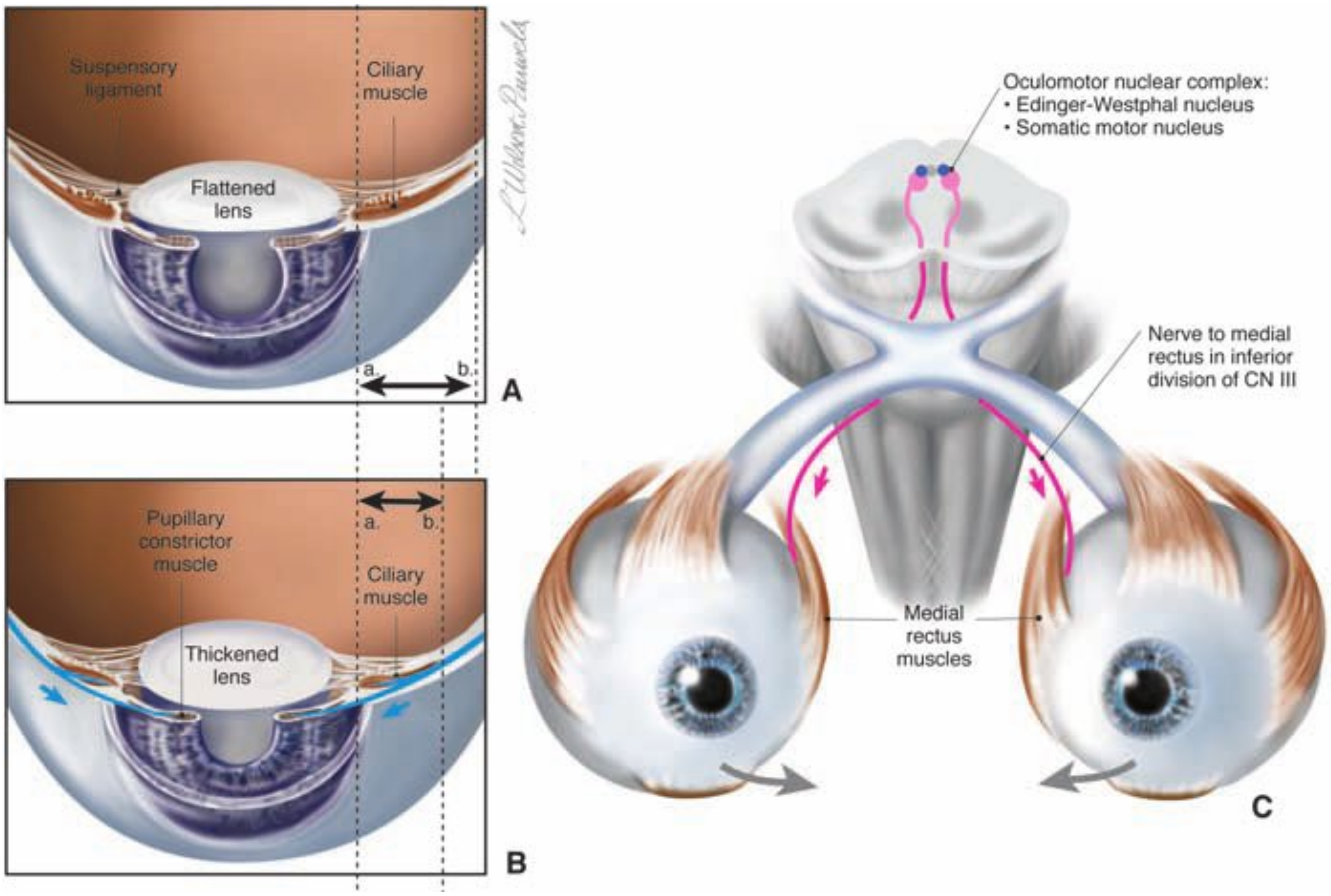
Suspensory ligament
Ciliary muscle
Flattened lens
a.
b.
A
Pupillary constrictor muscle
a.
b.
Ciliary muscle
Thickened lens
B
Oculomotor nuclear complex:
• Edinger-Westphal nucleus
• Somatic motor nucleus
Nerve to medial rectus in inferior division of CN III
Medial rectus muscles
C

FIGURE III–10 **A**, Eye adjusted for distance vision: large pupil and relaxed ciliary muscle. **B**, In accommodation for near vision, the pupillary constrictor muscles contract resulting in a smaller pupil, and the ciliary muscles contract and the suspensory ligaments relax resulting in a thicker lens. **C**, The medial recti muscles contract causing the eyes to converge.

- **An increase in the curvature of the lens.** The suspensory ligament of the lens is attached to the lens periphery. At rest, the ligament maintains tension on the lens, keeping it relatively flat (see Fig. III–10A). During accommodation, motor signals from the Edinger-Westphal nucleus cause the ciliary muscle to contract, shortening the distance from "a" to "b," thereby releasing some of the tension of the suspensory ligament of the lens and allowing the curvature of the lens to increase (see Fig. III–10B).
- **Pupillary constriction.** The Edinger-Westphal nucleus also signals the sphincter-like pupillary constrictor muscle to contract. The resulting smaller pupil helps to sharpen the image on the retina (see Fig. III–10B).
- **Convergence of the eyes.** The oculomotor nucleus sends signals to both medial rectus muscles, which cause them to contract. This, in turn, causes the eyes to converge in order to keep the object centered on the foveas (see Fig. III–10C).

CASE HISTORY GUIDING QUESTIONS

1. How did a posterior communicating artery aneurysm cause Werner's symptoms?
2. When light was shone into Werner's eyes, why did the left pupil constrict but not the right?
3. What is the significance of the light sensitivity Werner experienced in the 2 weeks preceding the subarachnoid hemorrhage?
4. Where else along the course of CN III could damage occur?
5. How can a third nerve palsy caused by damage to the neuronal cell bodies in the oculomotor nucleus be differentiated from a third nerve palsy caused by damage to the axons within the nerve itself?
6. Why did Werner's right eye deviate downward and outward?

1. How did a posterior communicating artery aneurysm cause Werner's symptoms?

The CN III passes close to the posterior communicating artery. An aneurysm (an expansion of the diameter of a blood vessel) of the posterior communicating artery can compress CN III, resulting in a lower motor neuron lesion (Fig. III–11). In Werner's case, the aneurysm was compressing the right CN III, causing his double vision and light sensitivity. Rupture of the aneurysm resulted in a subarachnoid hemorrhage, which caused his sudden, severe headache.

2. When light was shone into Werner's eyes, why did the left pupil constrict but not the right?

The aneurysm of the posterior communicating artery compressed the right oculomotor nerve but did not affect the optic nerve. When light was shone into the right eye, the afferent (special sensory) limb of the pupillary light response was intact. Light shone into either pupil causes signals to be sent along the optic nerve, which bilaterally innervates the Edinger-Westphal nuclei via the pretectal olivary nucleus. Visceral motor signals that arise from the Edinger-Westphal nuclei are transmitted along the parasympathetic fibers in CN III. In Werner's case, these fibers are intact on the left side but damaged on the right. Therefore, the left pupil constricted in response to both direct and indirect stimulation, but the right pupil did not constrict in either case (see Fig. III–11).

Compare Werner's *efferent* (*motor*) pupillary defect (see Fig. III–11) with Meredith's *afferent* (*sensory*) pupillary defect shown in Chapter II (Fig. II–12).

3. What is the significance of the light sensitivity Werner experienced in the 2 weeks preceding the subarachnoid hemorrhage?

The parasympathetic fibers responsible for constriction of the pupil in response to light lie on the superomedial surface of CN III (see Fig. III–2). Initially, the aneurysm was small and, therefore, only the parasympathetic fibers were compromised.

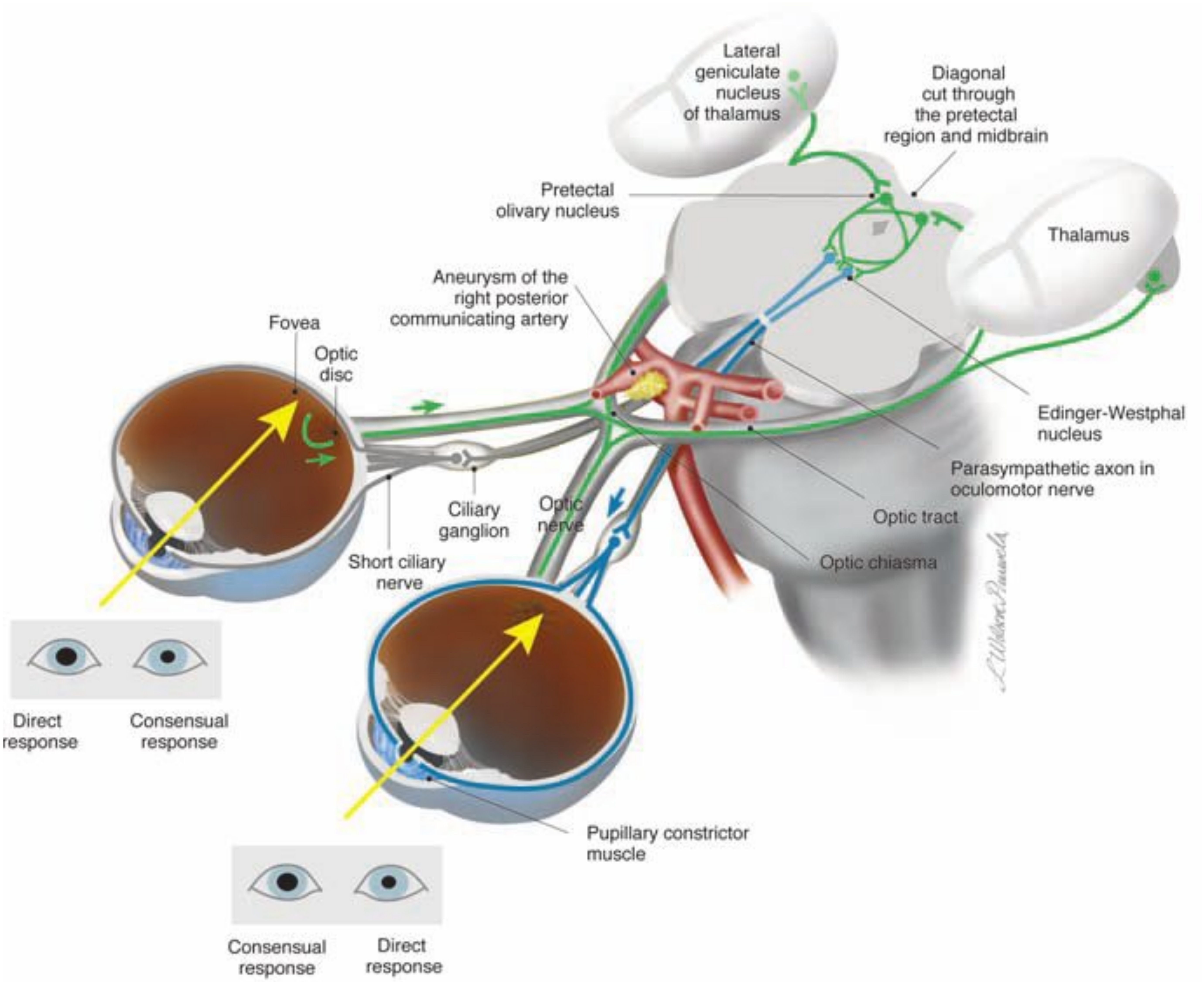
Lateral geniculate nucleus of thalamus
Diagonal cut through the pretectal region and midbrain
Pretectal olivary nucleus
Thalamus
Aneurysm of the right posterior communicating artery
Fovea
Optic disc
Edinger-Westphal nucleus
Parasympathetic axon in oculomotor nerve
Ciliary ganglion
Optic nerve
Optic tract
Short ciliary nerve
Optic chiasma
Direct response
Consensual response
Pupillary constrictor muscle
Consensual response
Direct response

FIGURE III–11 Efferent pupillary defect. Light shone in either of Werner's eyes resulted in left pupillary constriction (direct and consensual) but no constriction of the right pupil, due to an aneurysm of the posterior communicating artery.

As a result, Werner's right pupil could not constrict as well as usual in bright light, which caused his light sensitivity.

4. Where else along the course of CN III could damage occur?

CN III can be damaged anywhere from the nucleus in the midbrain to the muscles of innervation.

CN III can be damaged at the following sites:

Nucleus of CN III

- Although rare, damage to cells in the nucleus of CN III may be due to trauma, ischemia, or demyelination within the midbrain.

Peripheral Axons

- Damage to axons in the subarachnoid space may be due to infection, tumor infiltration, or infarction (loss of blood supply, usually caused by diabetes or hypertension).
- Compression of axons may be due to aneurysms, most typically in the posterior communicating artery and sometimes in the basilar artery (see Fig. III–11).
- Compression of axons may be caused by the uncus of the temporal lobe during cerebral herniation if there is raised intracranial pressure.
- Compression of axons in the cavernous sinus may be due to tumors, inflammation, infection, or thrombosis (other nerves that pass through the cavernous sinus [IV, V_1, V_2, VI] may also be involved) (see Fig. III–2).
- Damage may be caused by trauma to the area where axons pass through the superior orbital fissure to enter the orbit.

5. How can a third-nerve palsy caused by damage to the neuronal cell bodies in the oculomotor nucleus be differentiated from a third-nerve palsy caused by damage to the axons within the nerve itself?

To distinguish a nuclear lesion (see Fig. III–12A) from an axonal lesion (see Fig. III–12B), a useful indictor is the behavior of the upper eyelid.

In a nuclear lesion, there is

- No ptosis because there is bilateral innervation of the levator palpebrae superioris muscle by the central subnucleus.
- No weakness in upward gaze in the ipsilateral eye, but weakness of upgaze in the contralateral eye due to contralateral innervation of the superior rectus muscle.
- Ipsilateral weakness of downward gaze due to ipsilateral innervation of the inferior rectus muscle by the lateral subnucleus.
- Ipsilateral weakness of adduction due to ipsilateral innervation of the medial rectus muscle by the lateral subnucleus.
- Dilated unresponsive pupil due to ipsilateral innervation of the pupil by the Edinger-Westphal nucleus.

In contrast, when the peripheral part of the third nerve is damaged (axonal damage), innervation of all target muscles is affected and there is ptosis and a dilated unresponsive pupil on the ipsilateral side (see Fig. III–12B). Both lesions described previously are lower motor neuron lesions.

It is rare to have an exclusively unilateral oculomotor nucleus lesion because of its close proximity to the midline. Frequently, the central subnucleus, which resides in the midline, is bilaterally affected, resulting in bilateral ptosis.

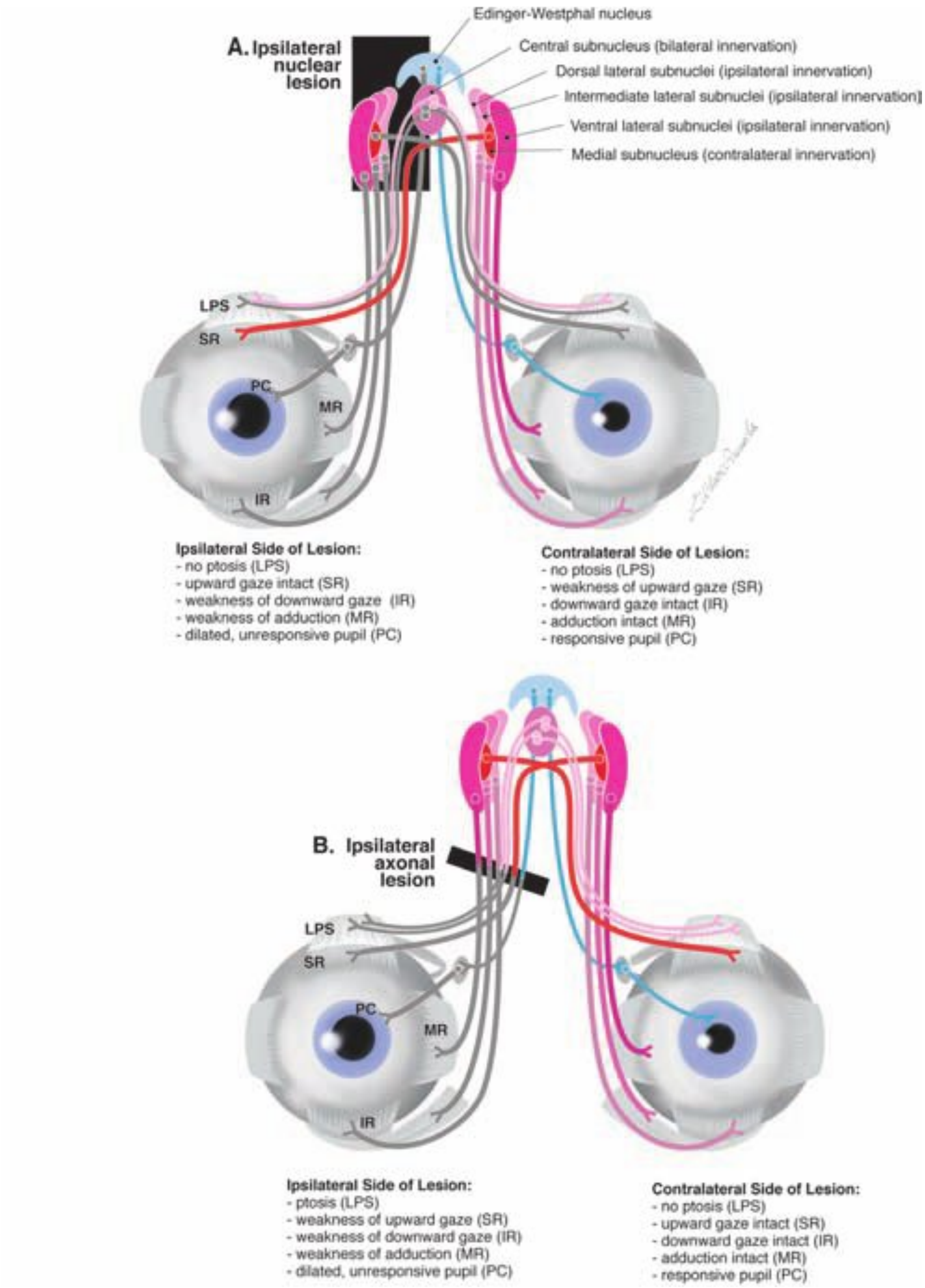
A. Ipsilateral nuclear lesion
Edinger-Westphal nucleus
Central subnucleus (bilateral innervation)
Dorsal lateral subnuclei (ipsilateral innervation)
Intermediate lateral subnuclei (ipsilateral innervation)
Ventral lateral subnuclei (ipsilateral innervation)
Medial subnucleus (contralateral innervation)
LPS
SR
PC
MR
IR
Ipsilateral Side of Lesion:
- no ptosis (LPS)
- upward gaze intact (SR)
- weakness of downward gaze (IR)
- weakness of adduction (MR)
- dilated, unresponsive pupil (PC)
Contralateral Side of Lesion:
- no ptosis (LPS)
- weakness of upward gaze (SR)
- downward gaze intact (IR)
- adduction intact (MR)
- responsive pupil (PC)
B. Ipsilateral axonal lesion
LPS
SR
PC
MR
IR
Ipsilateral Side of Lesion:
- ptosis (LPS)
- weakness of upward gaze (SR)
- weakness of downward gaze (IR)
- weakness of adduction (MR)
- dilated, unresponsive pupil (PC)
Contralateral Side of Lesion:
- no ptosis (LPS)
- upward gaze intact (SR)
- downward gaze intact (IR)
- adduction intact (MR)
- responsive pupil (PC)

FIGURE III–12

A. Right unilateral nuclear lower motor neuron lesion of cranial nerve III nucleus.

B. Right unilateral peripheral lower motor neuron lesion of cranial nerve III axons.

LPS	Levator Palpebrae Superions
SR	Superior Rectus Muscle
IR	Inferior Rectus Muscle
MR	Medial Rectus Muscle
PC	Pupillary Constrictor Muscle

6. Why did Werner's right eye deviate downward and outward?

Werner has a lower motor neuron lesion of the right CN III. His symptoms include the following:

- Strabismus (inability to direct both his eyes toward the same object) and consequent diplopia (double vision).
- Right-sided ptosis (lid droop) due to inactivation of the levator palpebrae superioris muscle and the subsequent unopposed action of the orbicularis oculi muscle. Werner tries to compensate for ptosis by contracting his frontalis muscle to raise his eyebrow and attached lid.
- Dilation of his right pupil due to decreased tone of the constrictor pupillae muscle.
- Downward abducted right eye position due to the unopposed action of his right superior oblique and lateral rectus muscles.
- Inability to accommodate with his right eye.

This combination of symptoms is called a "third-nerve palsy" (Fig. III–13).

CLINICAL TESTING

Testing of CN III involves the assessment of

- Eyelidp osition.
- Pupillary response to light.

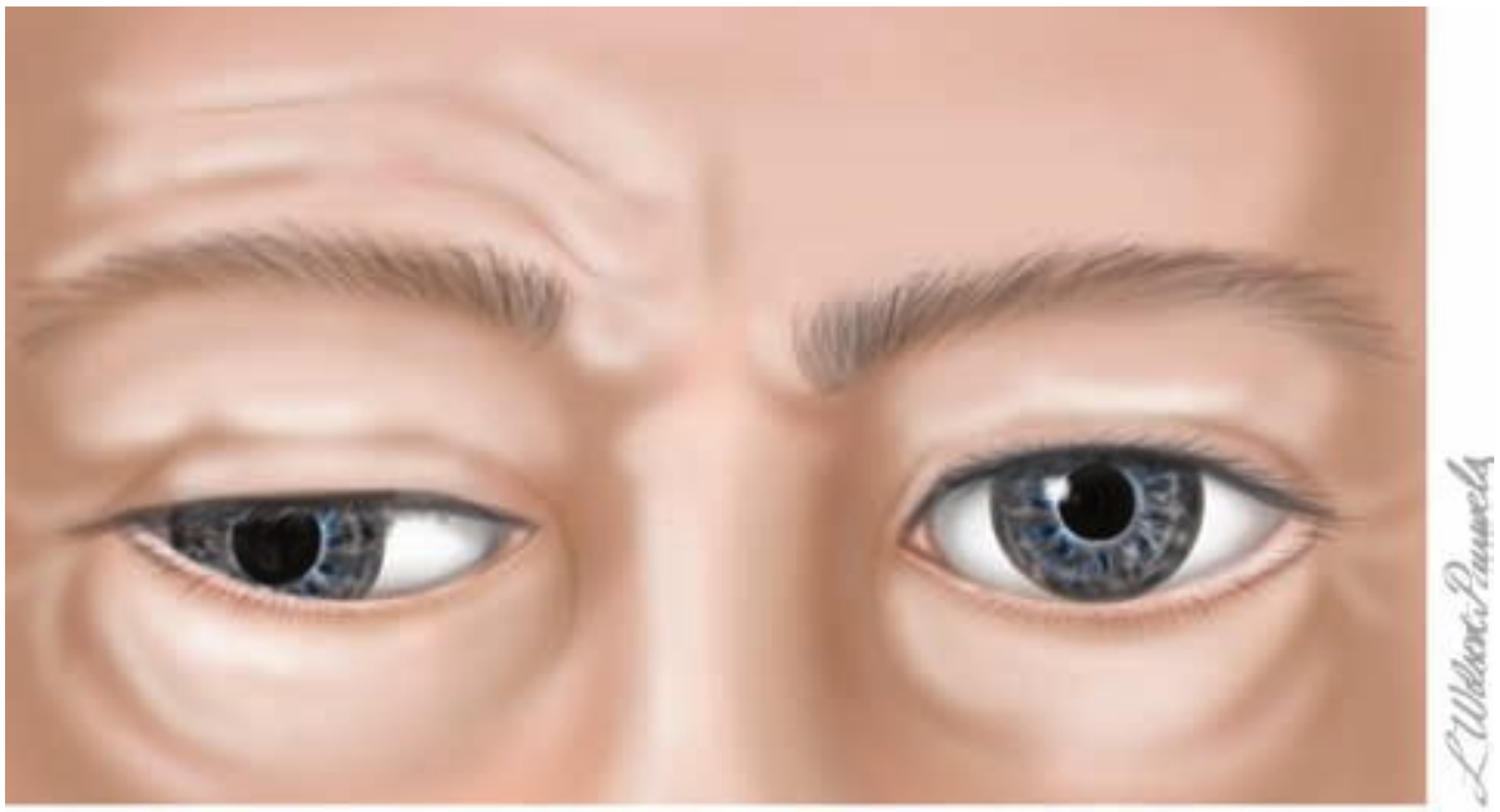

FIGURE III–13 Appearance of the eyes in right third nerve palsy.
The right side of Werner's face illustrates: a wrinkled brow due to the inability to raise the right eyelid; ptosis of his right eye lid due to the inactivation of the levator palpebrae superioris muscle; dilation of his right pupil due to the decreased tone of the constrictor pupillae muscle; and downward and outward movement of his right eye due to the unopposed action of the right superior oblique and lateral rectus muscles.

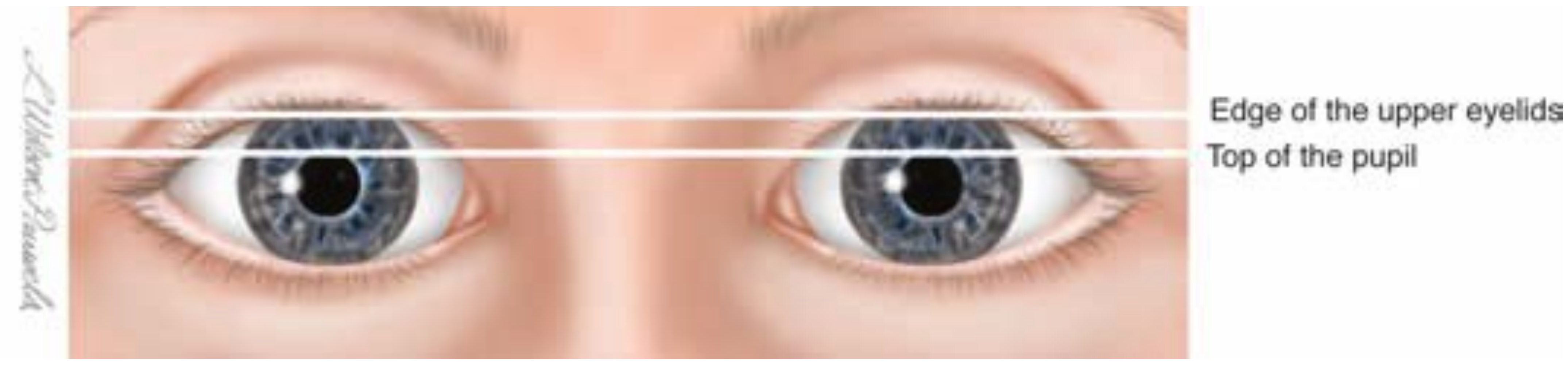

FIGURE III–14 Normal eyelid position.

- Accommodation.
- Extraocular eye movements.

(See also "Cranial Nerves Examination" on the web site.)

Eyelid Position

Elevation of the eyelid results from activation of the levator palpebrae superioris muscle. Damage to CN III will result in ipsilateral ptosis (drooping of the eyelid). To assess the function of the muscle, ask the patient to look directly ahead and note the position of the edge of the upper eyelid relative to the iris. The eyelid should not droop over the pupil, and the eyelid position should be symmetrical on both sides (Fig. III–14).

Pupillary Response to Light

The sensory limb of the pupillary light reflex is carried by CN II—the optic nerve (Fig. III–15). The motor component of the pupillary light reflex is carried by parasympathetic fibers that travel on the surface of CN III. To assess the integrity of the reflex, a beam of focused light is shone directly into one pupil. In the normal reflex, light shone in the ipsilateral eye causes the ipsilateral pupil to constrict (direct light response). The contralateral pupil also constricts (indirect or consensual response). If the sensory arm (CN II) is intact but the ipsilateral parasympathetic motor pathway is damaged, the ipsilateral pupil will not constrict but the contralateral pupil will (see Fig. III–11).

Accommodation

Accommodation allows the visual apparatus of the eye to focus on a near object. The observable events in accommodation are convergence and pupillary constriction of both eyes. Accommodation is tested by asking the patient to follow the examiner's finger as it is brought from a distance toward the patient's nose. As the examiner's finger approaches the patient's nose, the patient's eyes converge and her or his pupils constrict (Fig. III–16).

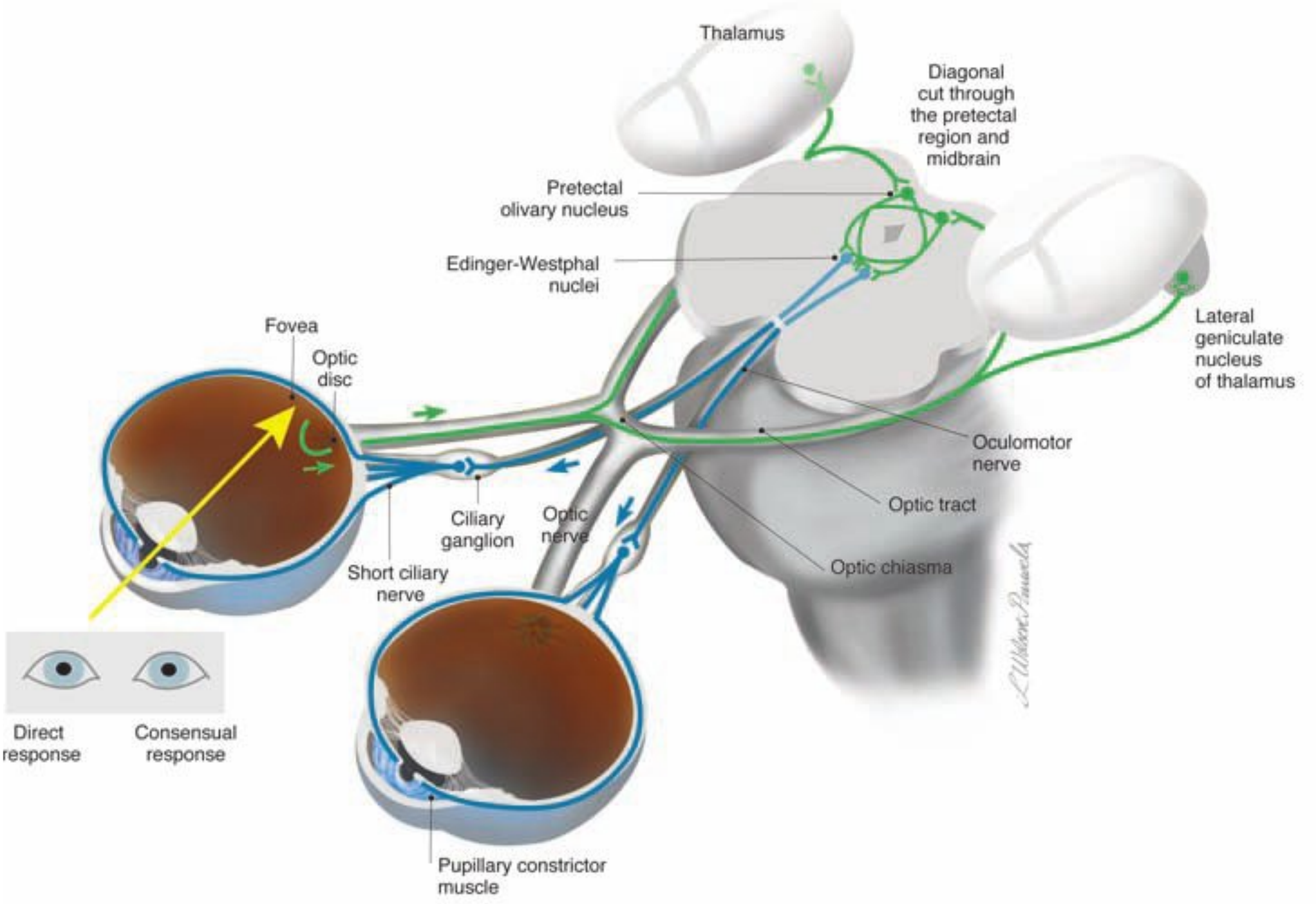

FIGURE III–15 Pupillary light reflex. Light shone in the right eye elicits pupillary constriction in the same eye (direct response) and in the opposite eye (consensual response).

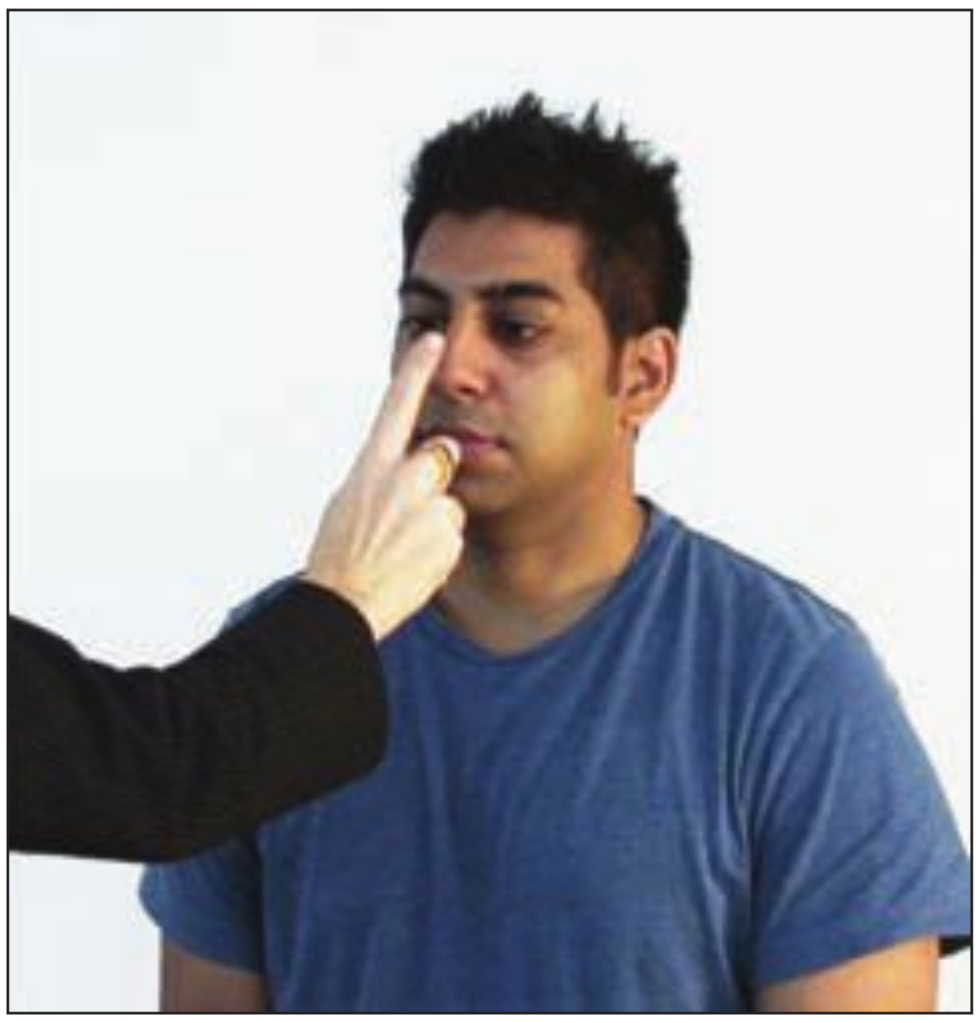

FIGURE III–16 Testing cranial nerve III for accommodation.

Extraocular Eye Movements

CN III is tested in conjunction with CNs IV and VI through assessment of extraocular eye movements. See Chapter XIII for a detailed description of extraocular assessment.

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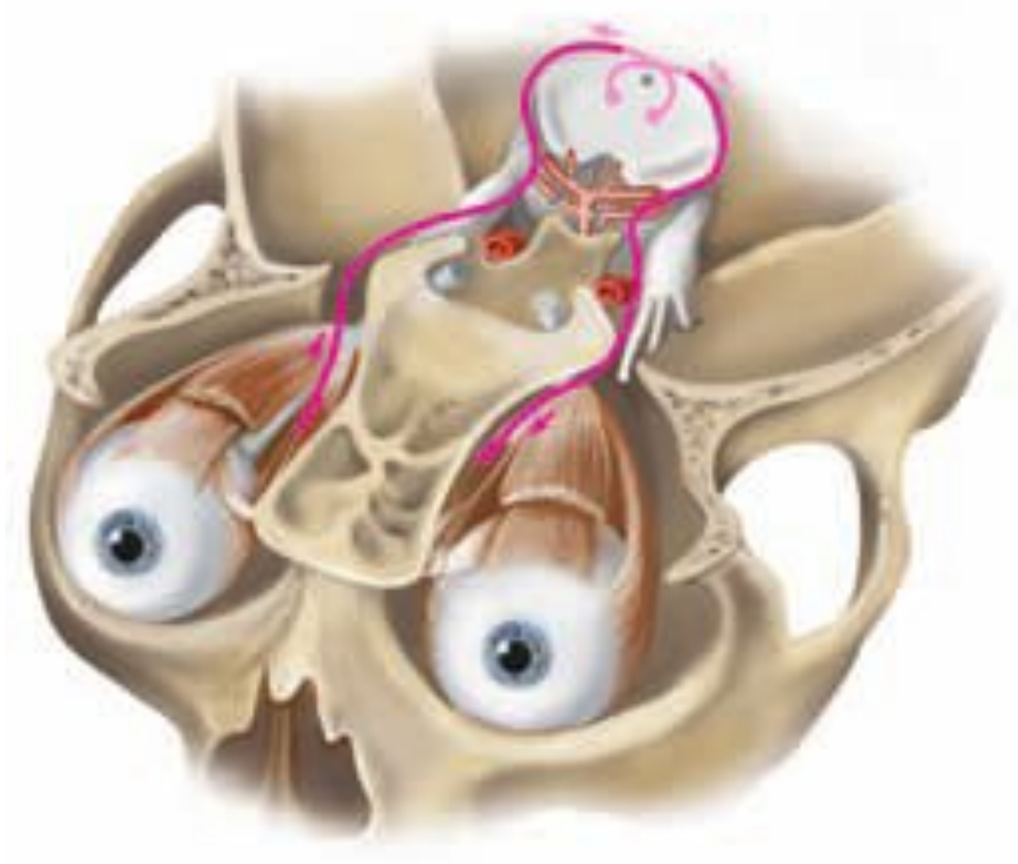


IV

Trochlear Nerve

CASE HISTORY

Lakshmi is a 53-year-old woman with a history of long-standing diabetes and hypertension. She was out for an afternoon walk when she experienced the onset of headache behind her right eye. It was a continuous dull pain and, although not severe, caused her to cut her walk short. While walking home, she experienced intermittent diplopia (double vision). Lakshmi assumed that she was just tired and expected it to pass. As she began to prepare dinner, she noticed that her diplopia worsened when she was looking down at the chopping board, but it improved if she tilted her head to the left or if she looked up. Alarmed that something was wrong, Lakshmi went to the hospital.

Initially, Lakshmi was assessed by a resident. The resident found that Lakshmi's eye movements were normal, and he could not account for her diplopia. Review of the case with a staff neurologist gave some clues to her problem. Lakshmi's pupils were equal in size and reactive to light, and there was no evidence of ptosis (upper eyelid droop). Lakshmi was able to move her eyes fully through the horizontal planes. However, when the neurologist tested Lakshmi's vertical eye movements, with her eyes abducted and adducted, he discovered that Lakshmi could not look down with her right eye when it was adducted. Lakshmi also noticed that she had diplopia in this position and that the image from her right eye was slightly lower than the image from her left eye. The neurologist diagnosed right cranial nerve IV palsy and attributed it to infarction of the nerve due to diabetes.

ANATOMY OF THE TROCHLEAR NERVE

The trochlear nerve (CN IV), the smallest of the cranial nerves, innervates a single muscle in the orbit: the superior oblique muscle (Figure IV–1A). It has only a somatic motor component (Table IV–1). The cell bodies of the lower motor neurons constitute the trochlear nucleus, which is located in the tegmentum of the midbrain at the level of the inferior colliculus (Figure IV–1B). Like other somatic

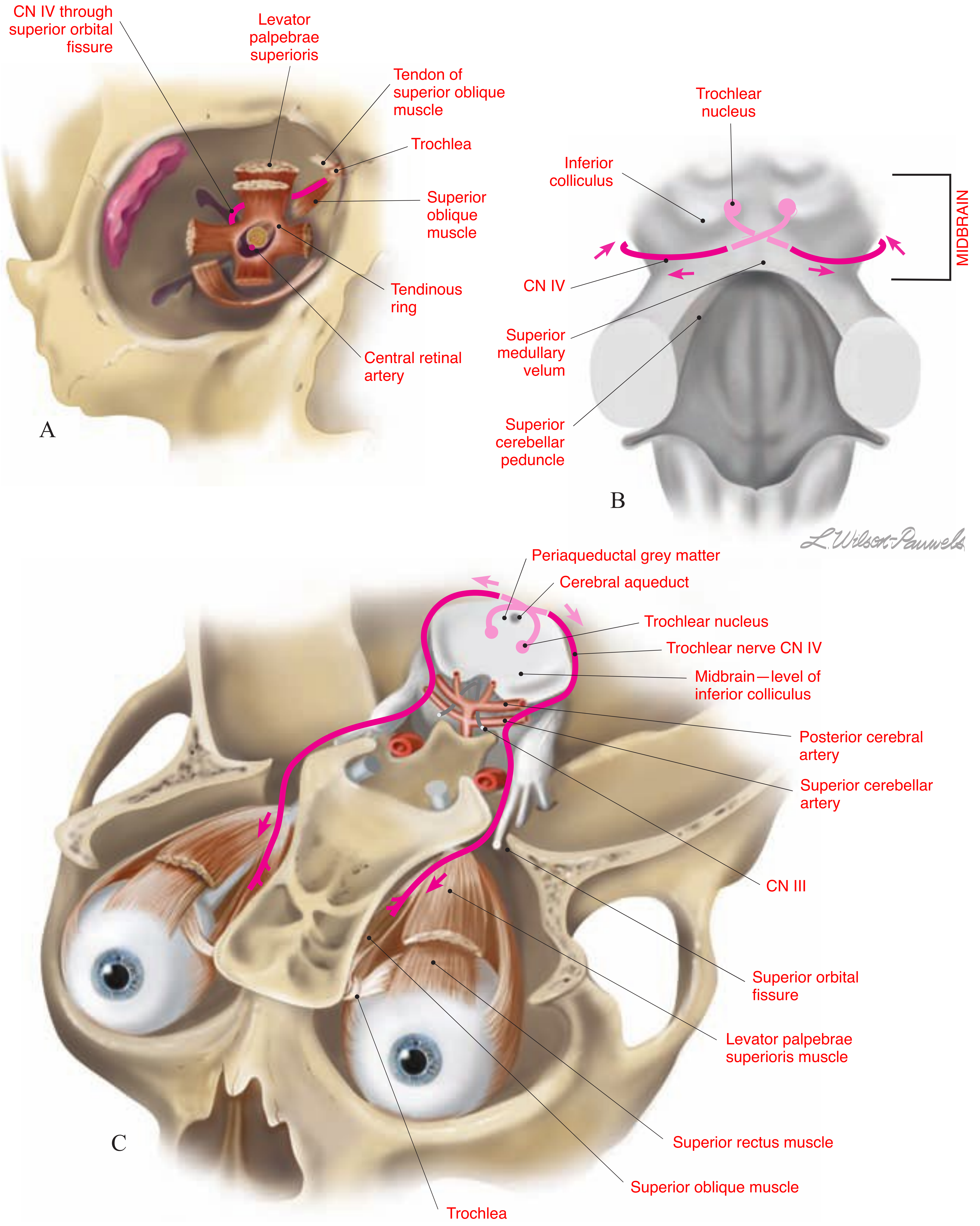


FIGURE IV–1 **A**, Apex of the right orbit illustrating the tendinous ring. **B**, Dorsal aspect of the brain stem. **C**, Somatic motor tracts from the trochlear nuclei in the brain stem to the superior oblique muscles.

motor nuclei, the trochlear nucleus is close to the midline. Motor neurons from the trochlear nucleus innervate predominantly, if not exclusively, the contralateral superior oblique muscle. Axons arising from the trochlear nucleus course dorsally around the periaqueductal grey matter and cerebral aqueduct, and cross the midline (Figure IV–1C). The crossed axons emerge from the dorsal aspect of the midbrain just caudal to the inferior colliculus to form cranial nerve IV. The nerve curves ventrally around the cerebral peduncle to pass between the posterior cerebral and superior cerebellar arteries lateral to cranial nerve III. Cranial nerve IV runs anteriorly to pierce the dura at the angle between the free and attached borders of the tentorium cerebelli.

TABLE IV–1 Component, Nucleus, and Function of the Trochlear Nerve

Component	Nucleus	Function
Somatic motor (efferent)	Trochlear Nucleus	Innervation of the superior oblique muscle of the eye

Cranial nerve IV enters the cavernous sinus along with cranial nerves III, V_1, V_2, and VI. Within the cavernous sinus, the trochlear nerve is situated between cranial nerves III and V_1 and lateral to the internal carotid artery (Figure IV–2). It leaves the cavernous sinus and enters the orbit through the superior orbital fissure, above the tendinous ring (see Figure IV–1A). The nerve then courses medially, close to the roof of the orbit, and runs diagonally above the levator palpebrae superioris muscle to reach its target, the superior oblique muscle. Here the nerve divides into three or more branches that enter the superior oblique muscle along its proximal third.

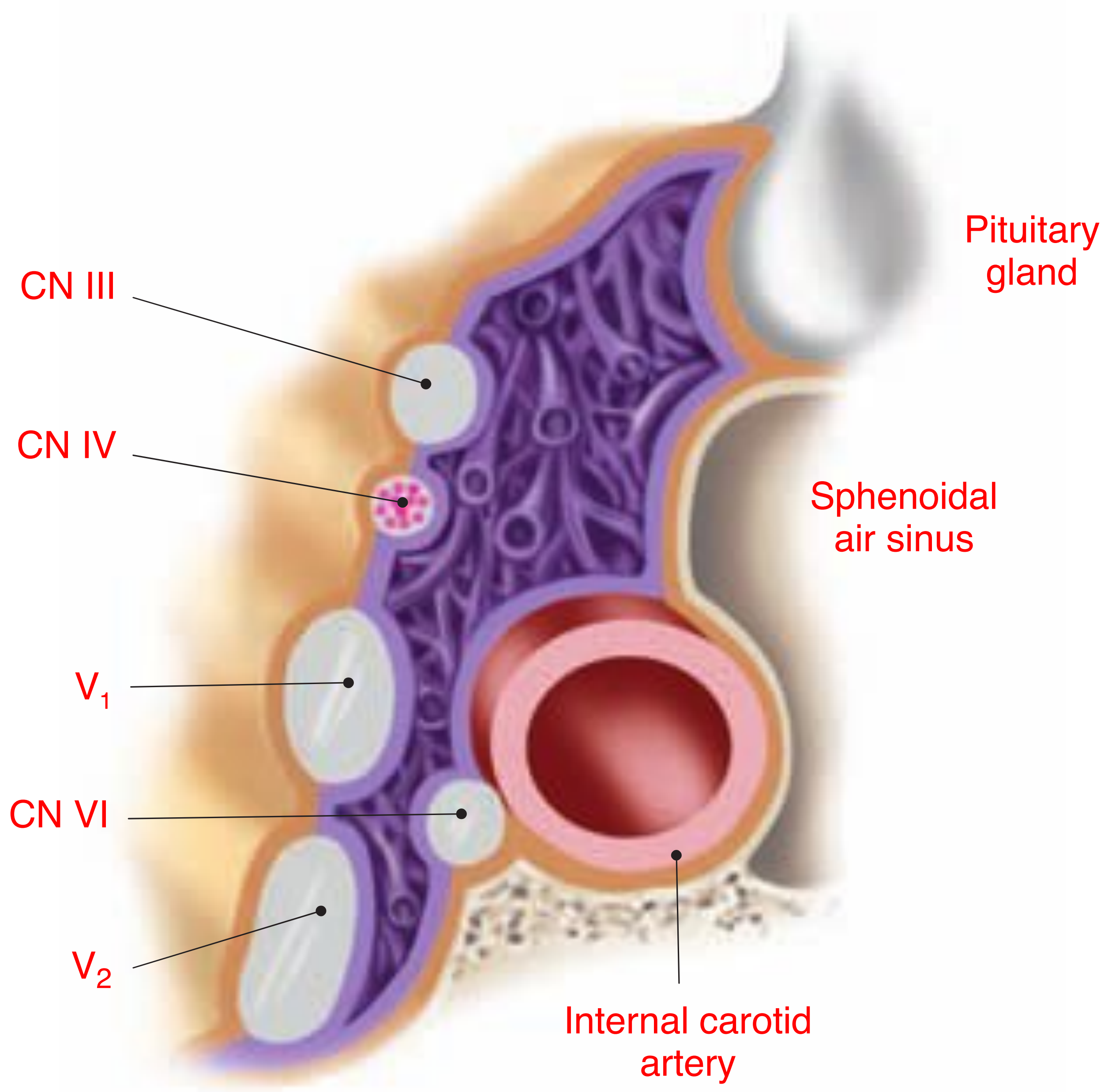
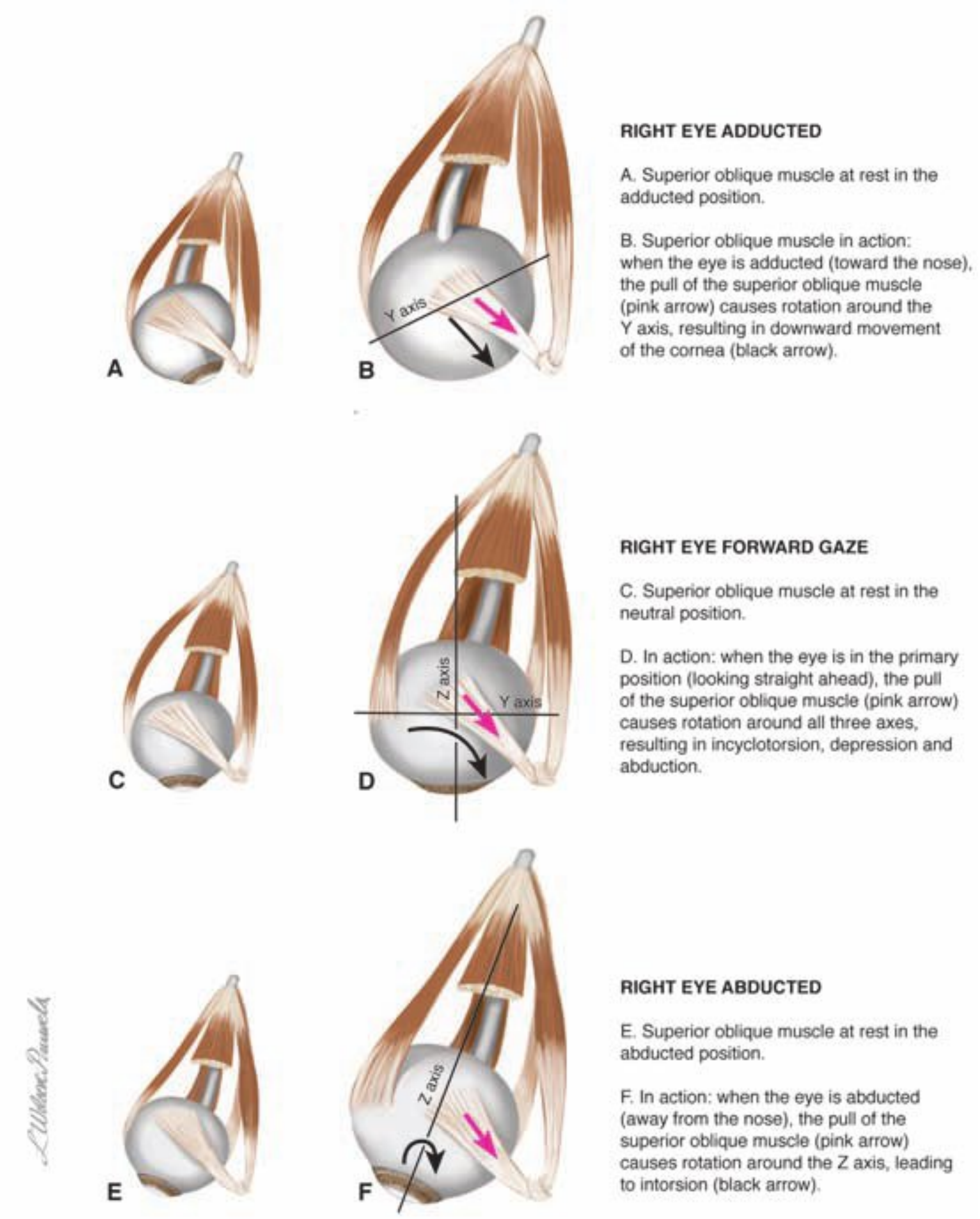


FIGURE IV–2 Slice through the right cavernous sinus showing the relationship of CN IV to other structures coursing through the sinus.

Eye Movements

From the tendinous ring and posterior roof of the orbit, the superior oblique muscle passes forward along the medial wall of the orbit to its superior anteromedial corner where its tendon loops through a "pulley" of fibrocartilage (the *trochlea*). It then turns posterolaterally to insert into the sclera in the superolateral posterior quadrant of the eye (Figure IV–3). Contraction of the muscle, therefore, pulls the posterolateral surface of the eyeball towards the trochlea.



RIGHT EYE ADDUCTED

A. Superior oblique muscle at rest in the adducted position.

B. Superior oblique muscle in action: when the eye is adducted (toward the nose), the pull of the superior oblique muscle (pink arrow) causes rotation around the Y axis, resulting in downward movement of the cornea (black arrow).

RIGHT EYE FORWARD GAZE

C. Superior oblique muscle at rest in the neutral position.

D. In action: when the eye is in the primary position (looking straight ahead), the pull of the superior oblique muscle (pink arrow) causes rotation around all three axes, resulting in incyclotorsion, depression and abduction.

RIGHT EYE ABDUCTED

E. Superior oblique muscle at rest in the abducted position.

F. In action: when the eye is abducted (away from the nose), the pull of the superior oblique muscle (pink arrow) causes rotation around the Z axis, leading to intorsion (black arrow).

FIGURE IV–3 Movements of the right eye produced by the action of the superior oblique muscle in different eye positions (viewed from above). For rotation around axes, see Figure IV–4.

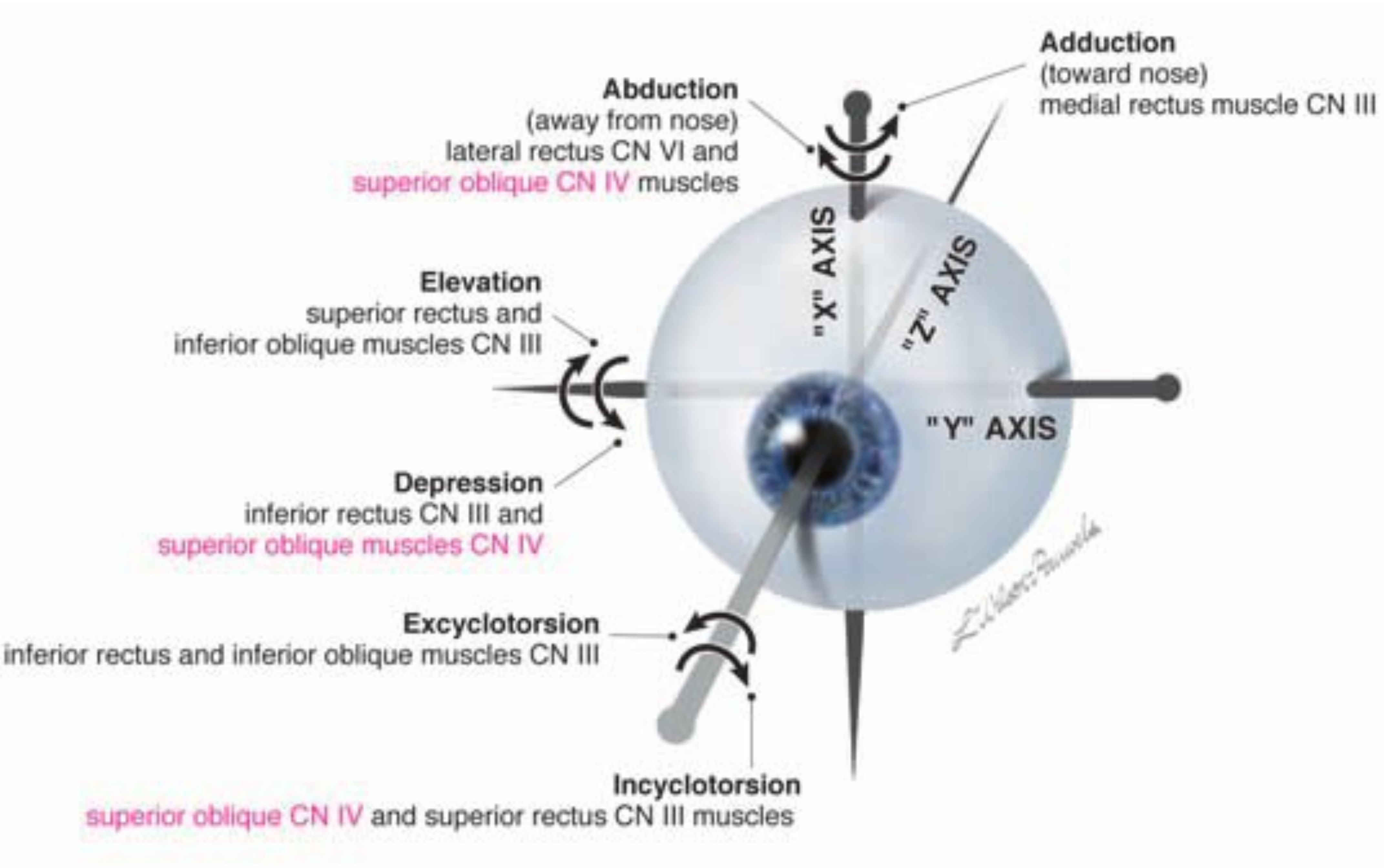

Abduction
(away from nose)
lateral rectus CN VI and
superior oblique CN IV muscles
Adduction
(toward nose)
medial rectus muscle CN III
"X" AXIS
"Z" AXIS
"Y" AXIS
Elevation
superior rectus and
inferior oblique muscles CN III
Depression
inferior rectus CN III and
superior oblique muscles CN IV
Excyclotorsion
inferior rectus and inferior oblique muscles CN III
Incyclotorsion
superior oblique CN IV and superior rectus CN III muscles

FIGURE IV–4 Right eye movements around the "X," "Y," and "Z" axes. Movements driven by CN IV are highlighted in pink.

The actual movement caused by the superior oblique muscle depends on the initial position of the eye (Table IV–2 and Figure IV–4).

TABLE IV–2 Eye Movements Mediated by Cranial Nerve IV

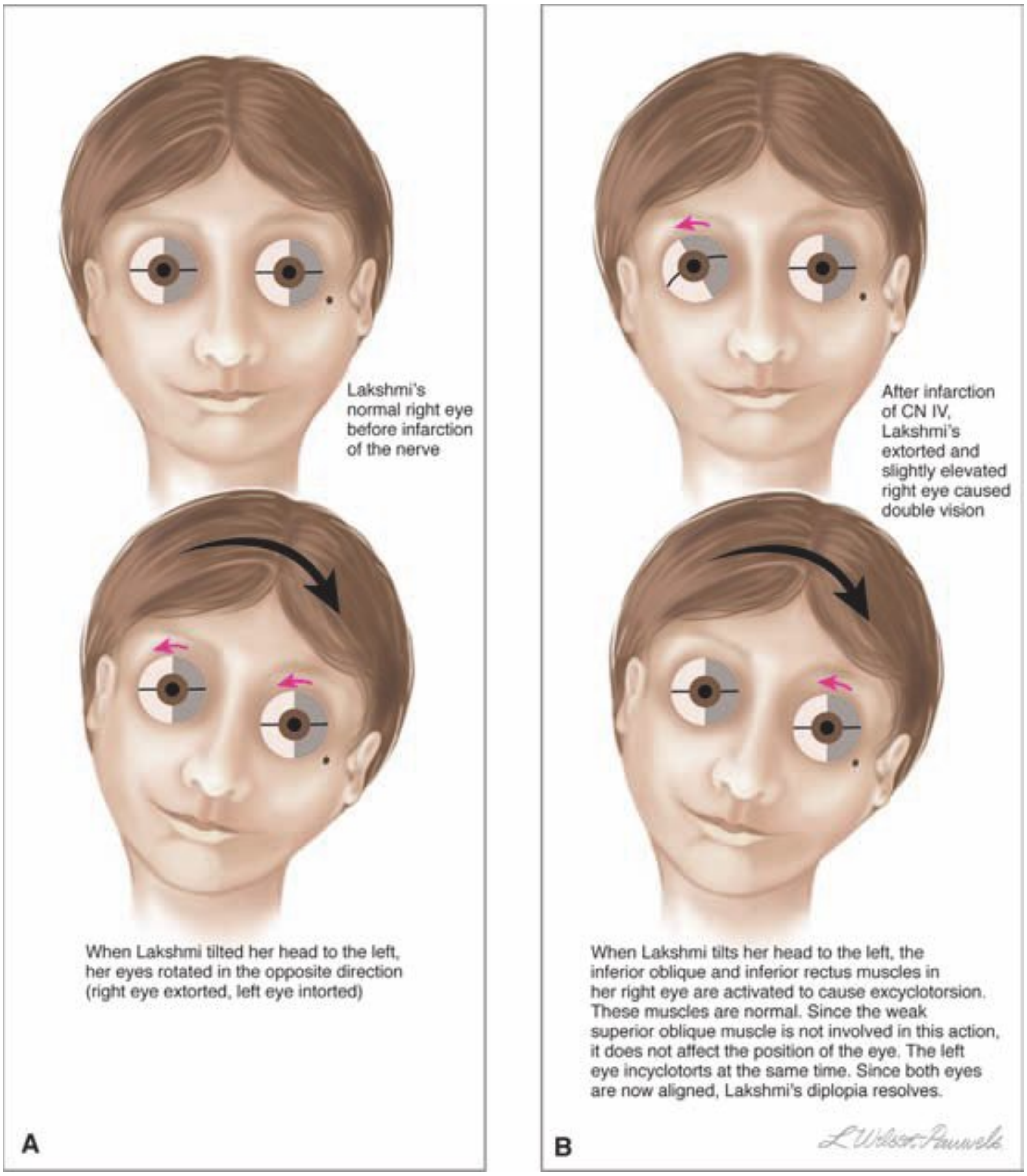
Nerve	Muscle	Primary Action	Subsidiary Action
CN IV	Superior oblique	Incyclotorsion	Depression and abduction

CASE HISTORY GUIDING QUESTIONS

1. Why did Lakshmi have vertical diplopia?
2. Why did Lakshmi's diplopia improve when she tilted her head to the left?
3. What other lesions could affect the trochlear nerve?
4. Why did the resident not identify Lakshmi's eye movement problem?

1. Why did Lakshmi have diplopia (double vision)?

Lakshmi's right superior oblique muscle is not functioning. As a result, its antagonist muscle, the inferior oblique, extorts and slightly elevates her right eye (Figure IV–5). Consequently, the visual fields are projected onto different areas of Lakshmi's right and left retinae and she perceives two distinct images.



Lakshmi's normal right eye before infarction of the nerve

When Lakshmi tilted her head to the left, her eyes rotated in the opposite direction (right eye extorted, left eye intorted)

After infarction of CN IV, Lakshmi's extorted and slightly elevated right eye caused double vision

When Lakshmi tilts her head to the left, the inferior oblique and inferior rectus muscles in her right eye are activated to cause excyclotorsion. These muscles are normal. Since the weak superior oblique muscle is not involved in this action, it does not affect the position of the eye. The left eye incyclotorts at the same time. Since both eyes are now aligned, Lakshmi's diplopia resolves.

FIGURE IV–5 Ocular rotation. **A**, Before infarction of CN IV; **B**, after infarction of CN IV.

2. Why did Lakshmi's diplopia improve when she tilted her head to the left?

Normally, as the head tilts from side to side, the eyes rotate in the opposite direction (see Figure IV–5A). Because Lakshmi's right superior oblique muscle was paralyzed, her right eye was extorted and slightly elevated (see Figure IV–5B, upper figure). When she tilted her head to the left, her left eye incyclotorted. At the same time in the right eye, the inferior rectus and inferior oblique muscles were activated to cause excyclotorsion. Since all the active muscles were normal, there was no imbalance and her diplopia improved (See Figure IV-5B, lower figure).

Clinical Comment

In contrast, when Lakshmi tilts her head to the right, the right eye tries to compensate by incyclotorting, which activates the superior rectus and superior oblique muscles (which are the incyclotorters of the right eye). Because the superior oblique muscle is weak, the vertical action of the superior rectus muscle (to raise the eye) outperforms the depressing action of the superior oblique muscle, resulting in a worsening of the vertical misalignment (right hypertropia). At the same time, the excyclotorters of the left eye are activated, but the inferior rectus and inferior oblique muscles are in balance in that eye, so the misalignment is only manifested in the right eye.

3. What other lesions could affect the trochlear nerve?

The trochlear nerve can be damaged anywhere from its nucleus in the midbrain to its termination in the orbit. The intramedullary (i.e., within the brainstem) axons can be damaged in the midbrain by infarction, tumor, or demyelination. The nerve can be damaged in the subarachnoid space by trauma, tumor, and meningitis. Within the cavernous sinus, aneurysm of the internal carotid artery, cavernous sinus thrombosis, tumors, and inflammation can all damage cranial nerve IV. The nerve can also be affected by ischemia; this is most commonly seen in patients with diabetes or hypertension.

4. Why did the resident not identify Lakshmi's eye movement problem?

The superior olbique muscle has three actions: incyclotorsion (rotation around the "Z" axis), downward gaze (rotation around the "Y" axis) and abduction (rotaton around the "X" axis) (See Figure IV–4). Because the eye is radially symmetric, rotation about the "Z" axis is very difficult to detect, but watching the movement of the conjunctival vessels is sometimes helpful. A better test of the superior oblique muscle is to examine downward gaze when the eye is adducted (Figure IV–6). Because the resident did not test Lakshmi's vertical eye movements with her right eye adducted, he missed the abnormality.

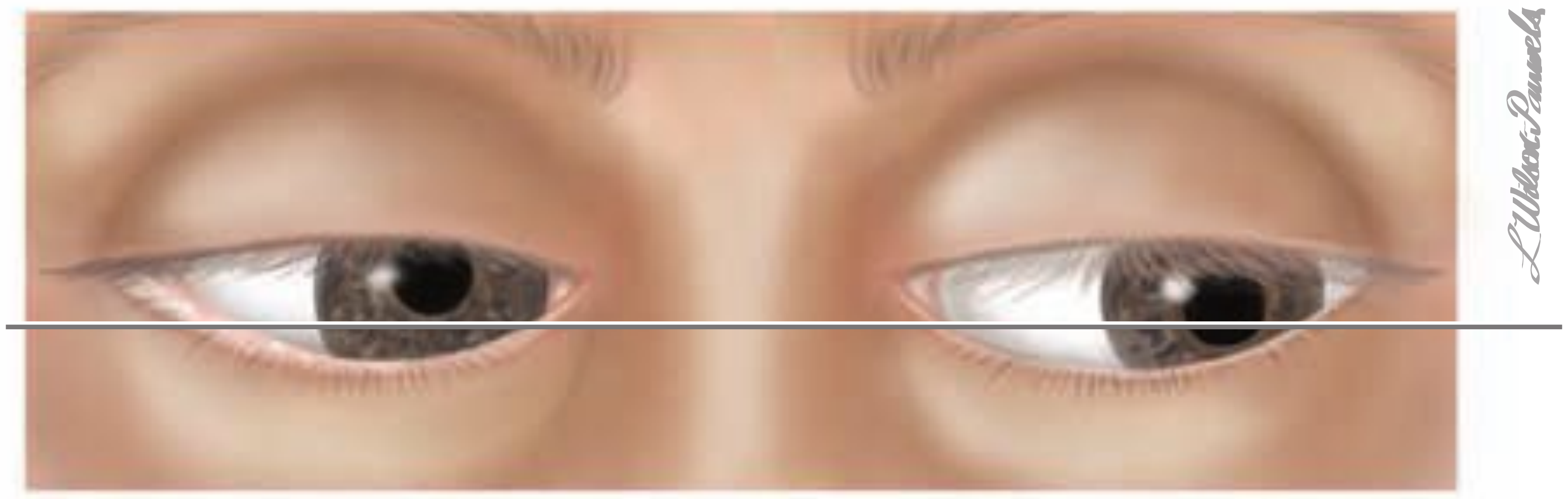


FIGURE IV–6 Lakshmi is unable to move her right eye downward when her right eye is adducted.

CLINICAL TESTING

Cranial nerve IV is tested in conjunction with cranial nerves III and VI through the assessment of eye movements (see Chapter 13).

ADDITIONAL RESOURCES

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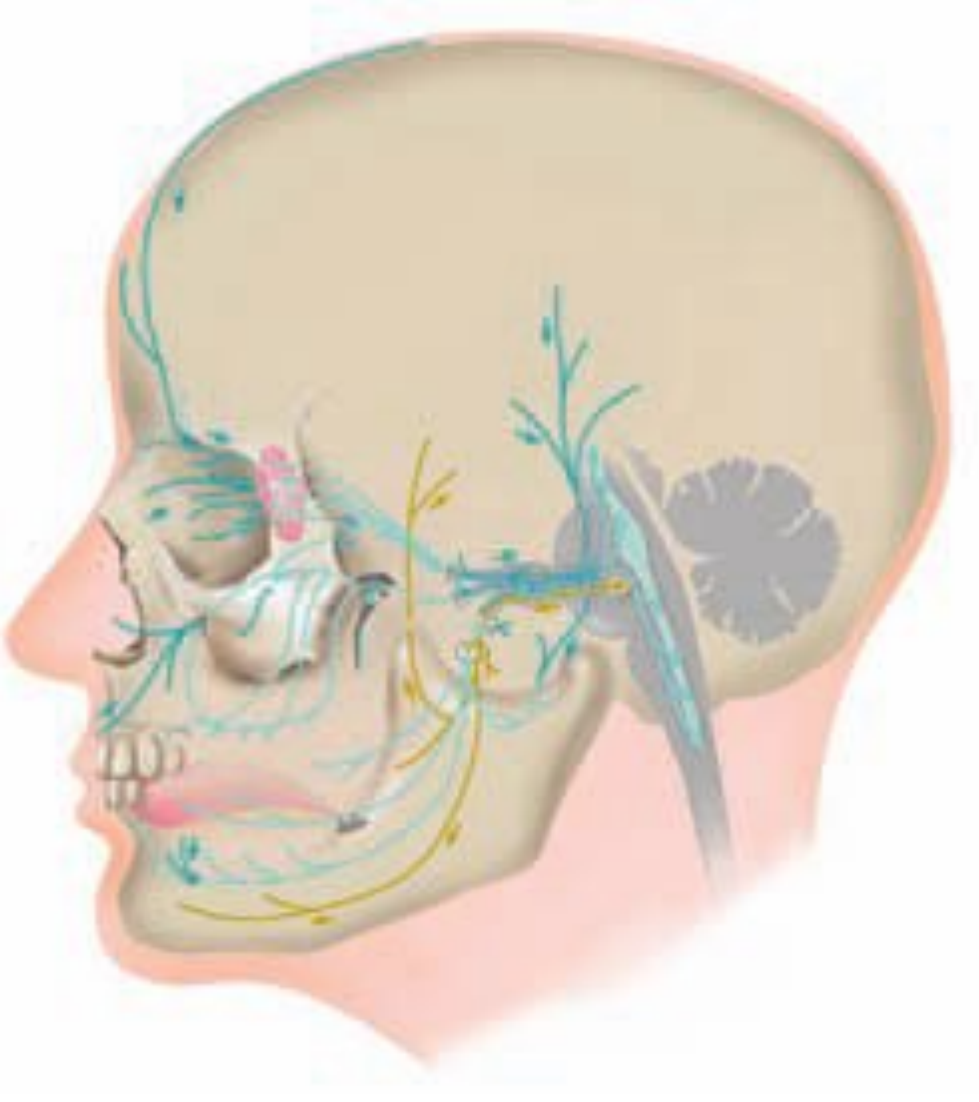

V

Trigeminal Nerve

CASE HISTORY

For the last few months, Mary, a 55-year-old professor, has been experiencing sudden attacks of extreme but short-lived pain on the left side of her face. She described the pain as "hornet stings" shooting through her jaw. The pain could be elicited by brushing her teeth, stroking the left side of her face, or talking. She initially thought her problem was related to her teeth and went to the dentist. Her dentist was unable to find anything wrong with her teeth and suggested that she see her family doctor.

When she saw her family doctor, Mary explained that the pain was most frequent when she was lecturing in class and had become so frequent and severe that she was finding it difficult to continue teaching. The doctor did a neurologic examination and found that her cranial nerves (CNs) were functioning normally. He suggested that her pain was probably due to hyperexcitablilty of her CN V (the trigeminal nerve) and diagnosed trigeminal neuralgia. To treat this condition, he prescribed carbamazepine. Initially, Mary's symptoms improved; however, months later, they returned and, despite trials with other anticonvulsants, the pain persisted. Because Mary was not improving, her doctor ordered magnetic resonance imaging (MRI) and referred her to a neurosurgeon for consideration of surgical treatment of her condition.

ANATOMY OF THE TRIGEMINAL NERVE

The name "trigeminal" (literally three born together) refers to the fact that CN V has three major divisions: the *ophthalmic* (V_1), *maxillary* (V_2), and *mandibular* (V_3) divisions (Figs. V–1 and V–2). It is the major sensory nerve of the face and innervates several muscles that are listed in Table V–1.

The trigeminal nerve emerges on the midlateral surface of the pons as a large sensory root and a smaller motor root. Its sensory ganglion (the semilunar or trigeminal

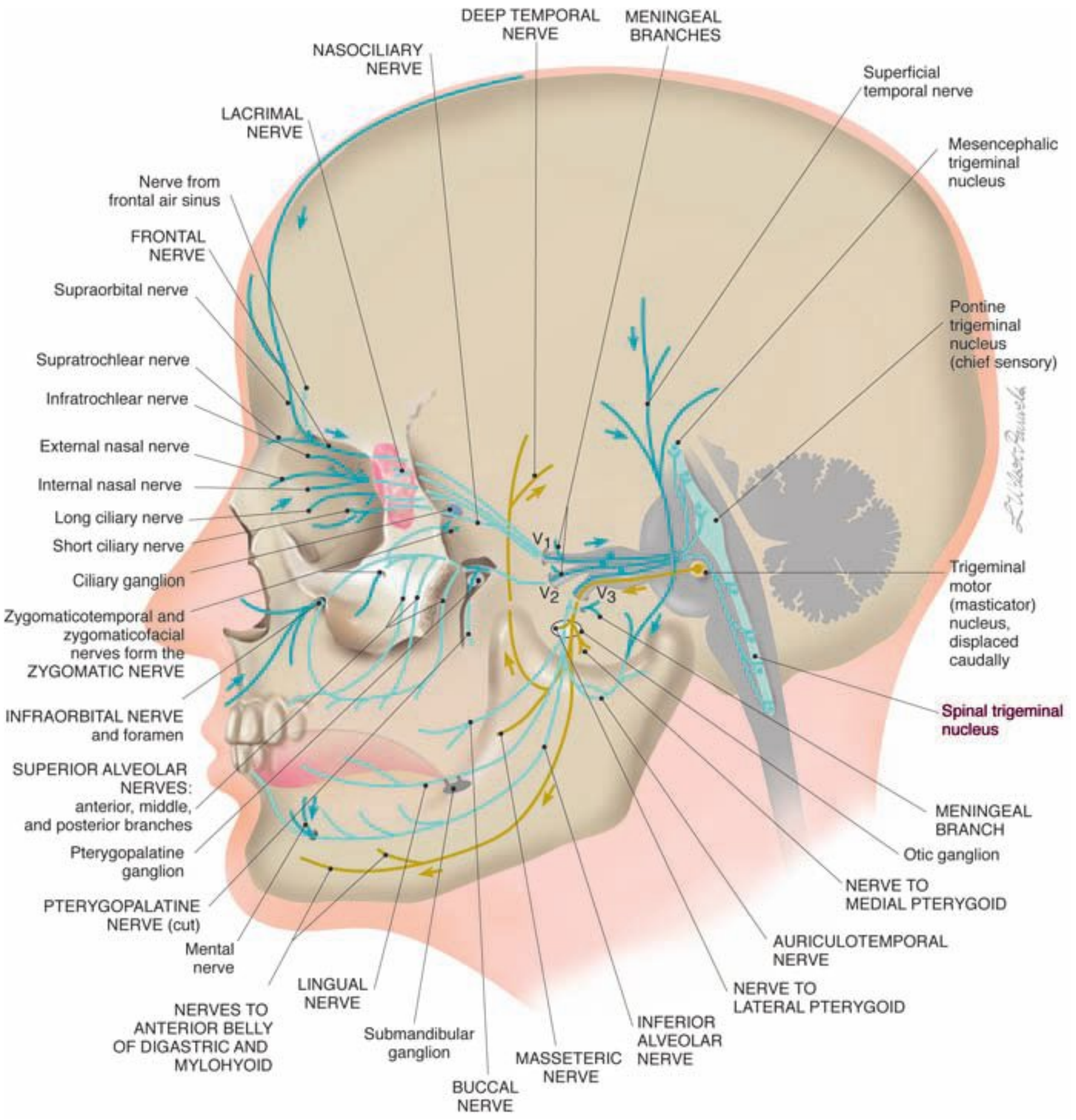
DEEP TEMPORAL NERVE
MENINGEAL BRANCHES
NASOCILIARY NERVE
LACRIMAL NERVE
Superficial temporal nerve
Mesencephalic trigeminal nucleus
Nerve from frontal air sinus
FRONTAL NERVE
Supraorbital nerve
Supratrochlear nerve
Infratrochlear nerve
External nasal nerve
Internal nasal nerve
Long ciliary nerve
Short ciliary nerve
Ciliary ganglion
Zygomaticotemporal and zygomaticofacial nerves form the ZYGOMATIC NERVE
INFRAORBITAL NERVE and foramen
SUPERIOR ALVEOLAR NERVES: anterior, middle, and posterior branches
Pterygopalatine ganglion
PTERYGOPALATINE NERVE (cut)
Mental nerve
NERVES TO ANTERIOR BELLY OF DIGASTRIC AND MYLOHYOID
LINGUAL NERVE
Submandibular ganglion
BUCCAL NERVE
MASSETERIC NERVE
INFERIOR ALVEOLAR NERVE
NERVE TO LATERAL PTERYGOID
AURICULOTEMPORAL NERVE
NERVE TO MEDIAL PTERYGOID
Otic ganglion
MENINGEAL BRANCH
Pontine trigeminal nucleus (chief sensory)
Trigeminal motor (masticator) nucleus, displaced caudally
Spinal trigeminal nucleus
V_1
V_2
V_3

FIGURE V–1 Overview of the trigeminal nerve.

or Gasserian ganglion) sits in a depression called the "trigeminal cave" (Meckel's cave) near the apex of the petrous partion of the temporal bone in the middle cranial fossa. Sensory axons at the distal aspect of the ganglion form the three major divisions (V_1, V_2, and V_3). The motor axons travel with the mandibular division (V_3).

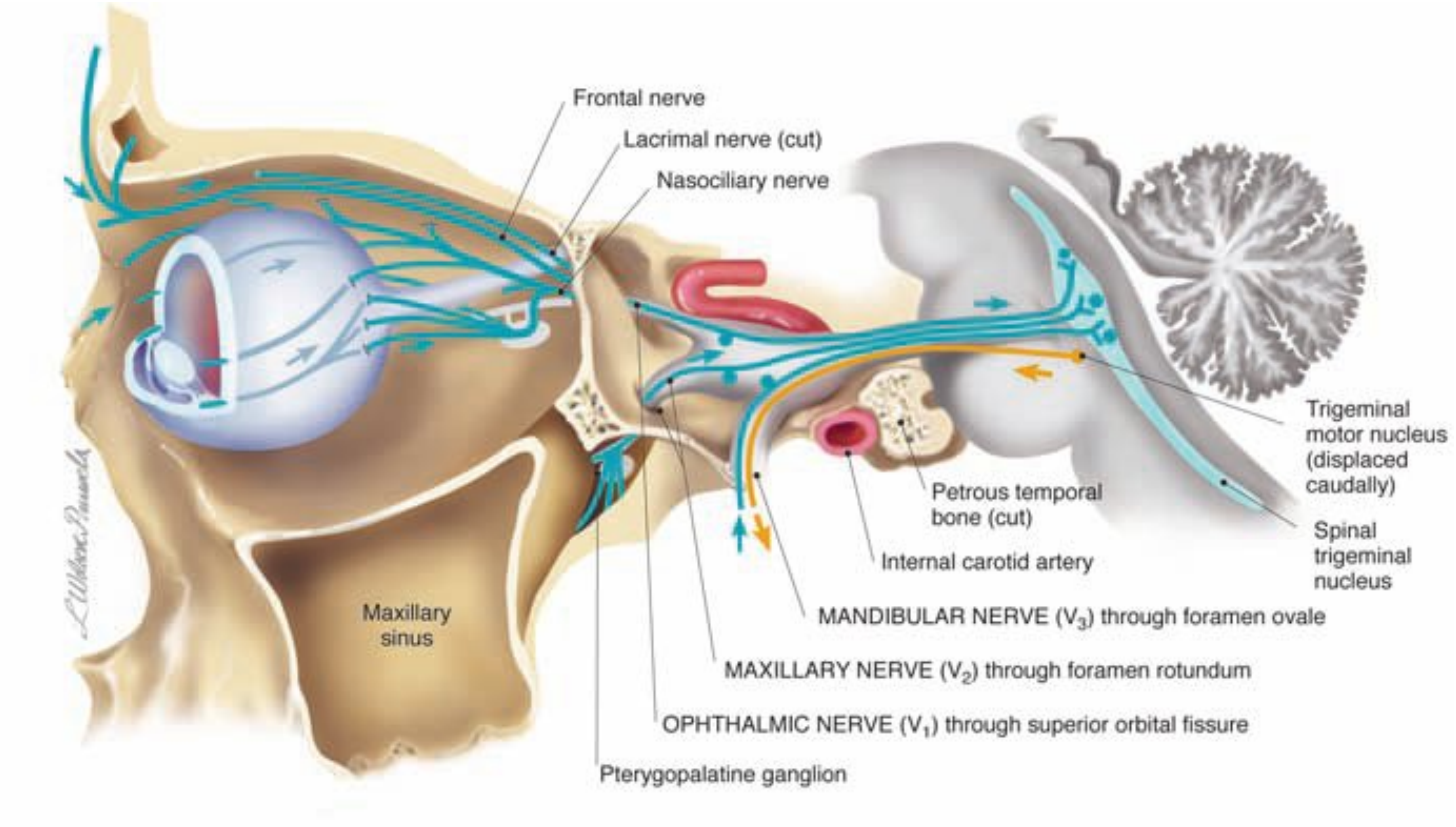
Frontal nerve
Lacrimal nerve (cut)
Nasociliary nerve
Trigeminal motor nucleus (displaced caudally)
Spinal trigeminal nucleus
Petrous temporal bone (cut)
Internal carotid artery
Maxillary sinus
MANDIBULAR NERVE (V_3) through foramen ovale
MAXILLARY NERVE (V_2) through foramen rotundum
OPHTHALMIC NERVE (V_1) through superior orbital fissure
Pterygopalatine ganglion

FIGURE V–2 Parasagittal section through the skull showing the trigeminal ganglion and its three major divisions. The motor (masticator) nucleus has been displaced caudally for illustrative purposes. It lies medial to the chief sensory nucleus.

TABLE V–1 Components, Nuclei, Ganglion and Functions of the Trigeminal Nerve (CN V)

Component	Nucleus	Ganglion	Function
General sensory (afferent)	Trigeminal nucleus	Trigeminal ganglion	For general sensation from the face and anterior scalp as far posteriorly as the apex of the head, conjunctivae, bulb of the eye, mucous membranes of paranasal sinuses, and nasal and oral cavities including the tongue and teeth, part of the external aspect of the tympanic membrane, and from the meninges of the anterior and middle cranial fossae*
Branchial motor (efferent)	Masticator nucleus		To innervate the muscles of mastication: masseter, temporalis, medial and lateral pterygoid muscles plus tensores tympani, tensores veli palatini, mylohyoid, and anterior belly of the digastric muscles

*Meninges of the posterior cranial fossa receive their sensory innervation from the upper few cervical nerves.

GENERAL SENSORY (AFFERENT) COMPONENT

Ophthalmic Division (V_1)

The ophthalmic division has three major branches (Table V–2): the frontal, lacrimal, and nasociliary nerves (Figs. V–3 and V–4; see also Fig. V–2).

Table V–2 Branches of the Trigeminal Nerve

Division	General Sensory	Branchial Motor
Ophthalmic (V_1)	■ Lacrimal ■ Frontal - Supratrochlear - Supraorbital nerve from frontal air sinus ■ Nasociliary - Long and short ciliary - Infratrochlear - Ethmoidal Anterior Internal nasal (medial and lateral) External nasal Posterior ■ Meningeal branch (from the tentorium cerebelli)	
Maxillary (V_2)	■ Zygomatic - Zygomaticotemporal - Zygomaticofacial ■ Infraorbital - External nasal - Superior labial ■ Superior alveolar - Anterior - Middle - Posterior ■ Nasociliary ■ Palatine - Orbital - Greater and lesser palatine - Posterior superior nasal - Pharyngeal ■ Meningeal - Anterior cranial fossa - Middle cranial fossa	

(continued)

Division	General Sensory	Branchial Motor
Mandibular (V_3)	■ Buccal ■ Lingual ■ Inferior alveolar - Dental - Incisive - Mental ■ Auriculotemporal - Anterior auricular - External acoustic meatus - Temporomandibular joint - Superficial temporal ■ Meningeal (spinosus) - Anterior cranial fossa - Middle cranial fossa	■ Medial ptergoid - Tensor veli palatini - Tensor tympani ■ Lateral pterygoid ■ Masseteric ■ Deep temporal ■ Mylohyoid - Nerve to mylohyoid - Nerve to anterior belly of digastric

The *frontal nerve* is formed by the supraorbital nerve from the forehead and scalp and the supratrochlear nerve from the bridge of the nose, medial part of the upper eyelid, and medial forehead. A small sensory twig from the frontal air sinus joins the frontal nerve near the anterior part of the orbit.

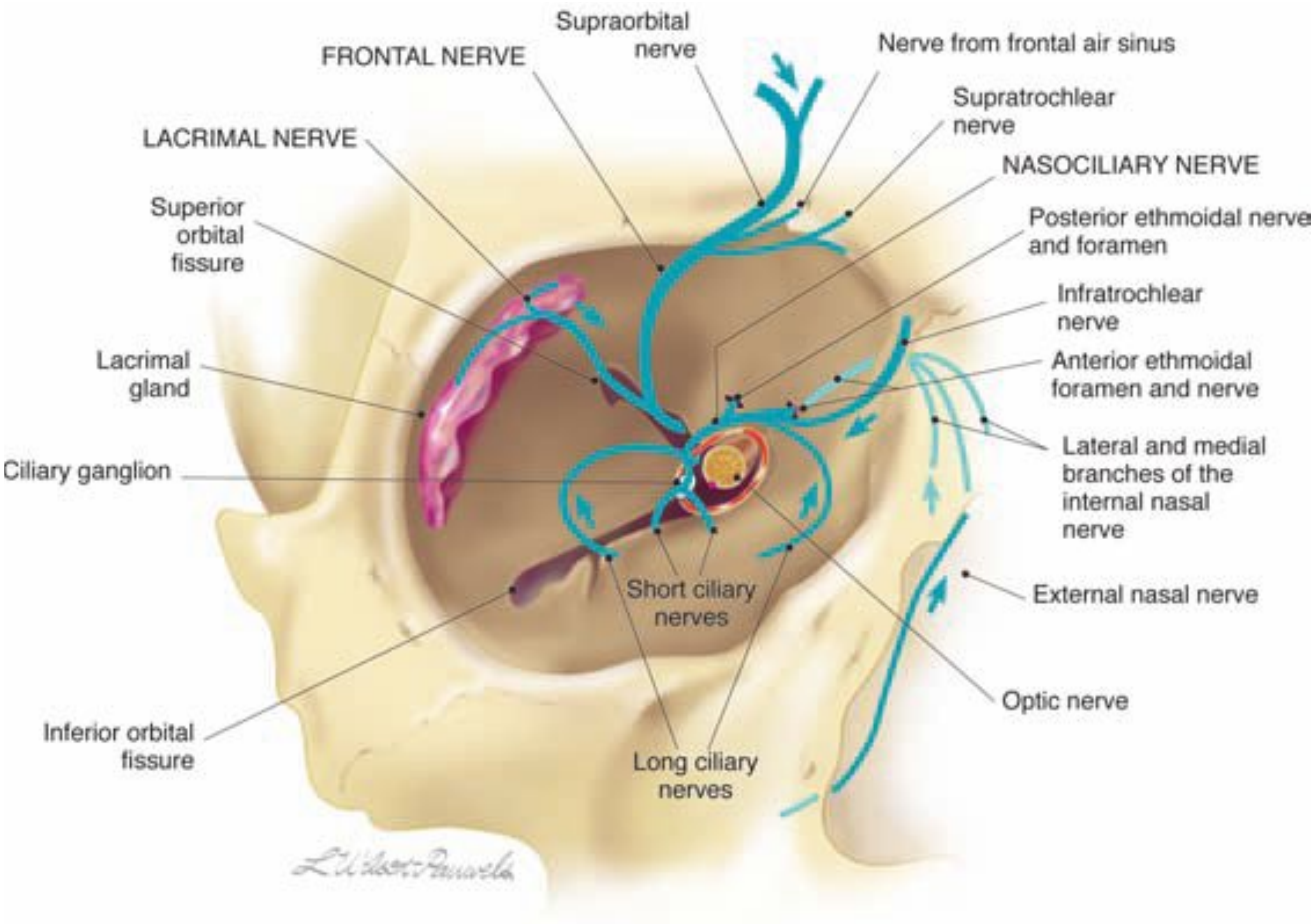

FIGURE V–3 Apex of right orbit illustrating branches of the ophthalmic division (V_1).

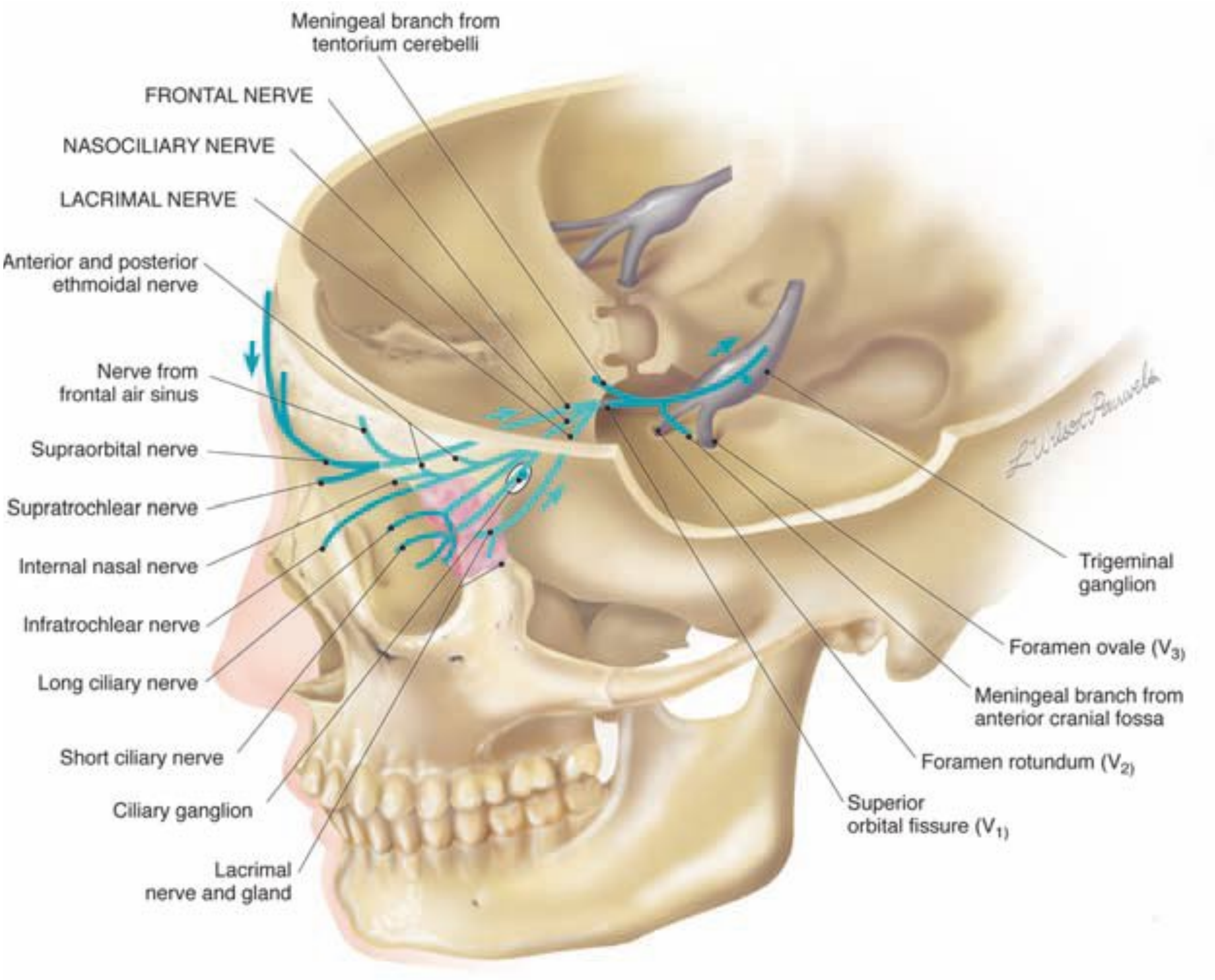

FIGURE V–4 General sensory component of the trigeminal nerve, ophthalmic V_1 division.

The *lacrimal nerve* carries sensory information from the lateral part of the upper eyelid, conjunctiva, and lacrimal gland. It runs posteriorly near the roof of the orbit to join the frontal and nasociliary nerves at the superior orbital fissure. Secretomotor fibers to the lacrimal gland from cranial nerve VII (facial) may travel briefly with the lacrimal nerve in its peripheral portion.

The *nasociliary nerve* is formed by the convergence of several terminal branches. These are the infratrochlear nerve from the skin of the medial part of the eyelid and side of the nose, the external nasal nerve from the skin of the ala and apex of the nose, the internal nasal nerve from the anterior part of the nasal septum and lateral wall of the nasal cavity, the anterior and posterior ethmoidal nerves from the ethmoidal air sinuses, and the long and short ciliary nerves from the bulb of the eye.

The ophthalmic division leaves the orbit through the superior orbital fissure, passes through the cavernous sinus, and enters the trigeminal ganglion. Here it is joined by a meningeal branch from the tentorium cerebelli.

Maxillary Division (V_2)

The maxillary division is formed by the zygomatic, infraorbital, superior alveolar, and palatine nerves (Figs. V–5 and V–6).

The *zygomatic nerve* has two major branches. Sensory processes from the prominence of the cheek converge to form the zygomaticofacial nerve. This nerve pierces the frontal process of the zygomatic bone and enters the orbit through its lateral wall. It turns posteriorly to join with the zygomaticotemporal nerve. Sensory processes from the side of the forehead converge to form the zygomaticotemporal nerve, which pierces the posterior aspect of the frontal process of the zygomatic bone and traverses the lateral wall of the orbit to join with the zygomaticofacial nerve, forming the zygomatic nerve. The zygomatic nerve courses posteriorly along the floor of the orbit to join with the maxillary nerve close to the inferior orbital fissure.

The *infraorbital nerve* is formed by cutaneous branches from the upper lip, medial cheek, and side of the nose. It passes through the infraorbital foramen of the

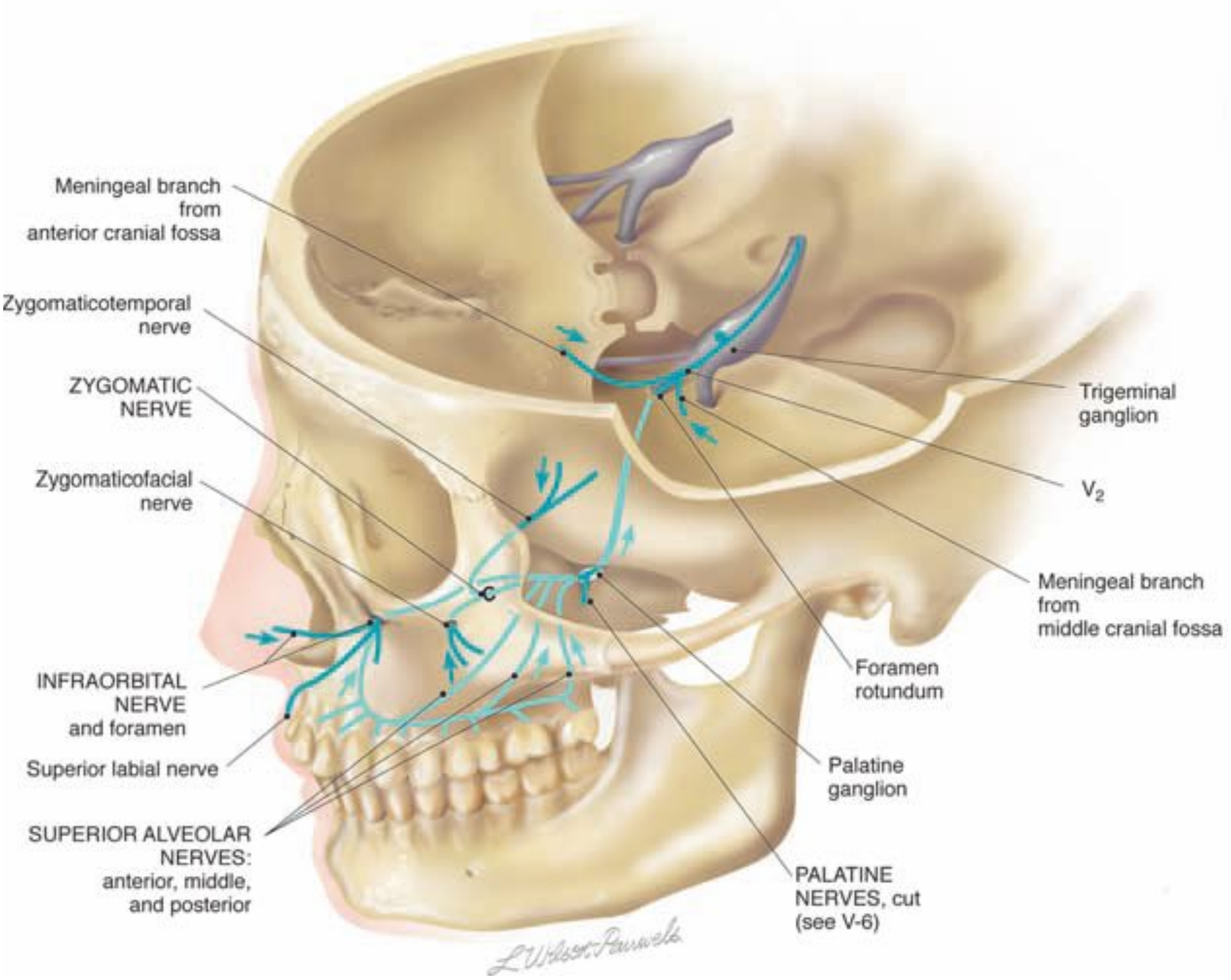
Meningeal branch from anterior cranial fossa
Zygomaticotemporal nerve
ZYGOMATIC NERVE
Zygomaticofacial nerve
C
INFRAORBITAL NERVE and foramen
Superior labial nerve
SUPERIOR ALVEOLAR NERVES: anterior, middle, and posterior
Trigeminal ganglion
V_2
Meningeal branch from middle cranial fossa
Foramen rotundum
Palatine ganglion
PALATINE NERVES, cut (see V-6)
L Wilson-Pauwels

FIGURE V–5 General sensory component of the trigeminal nerve - maxillary (V_2) division.

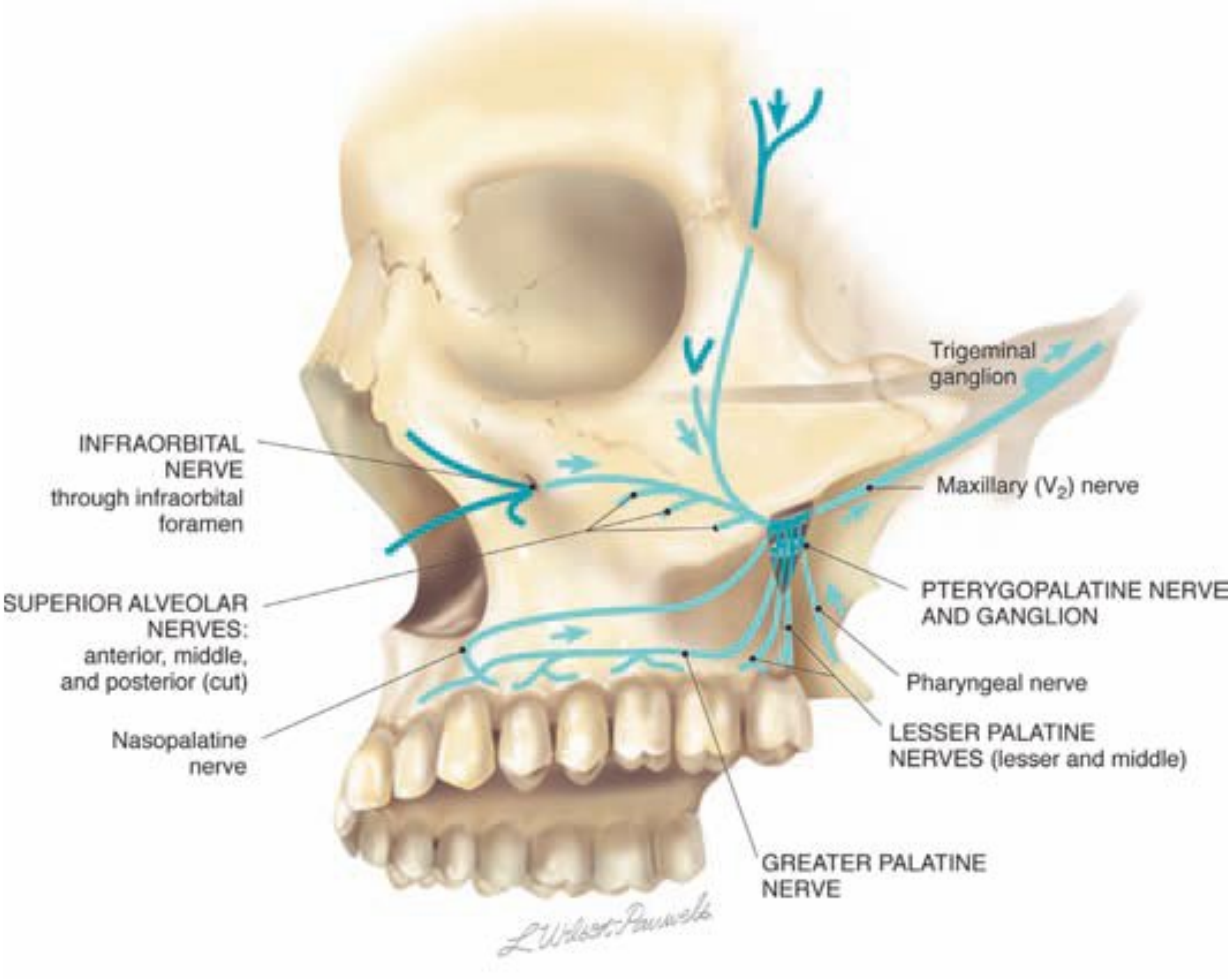
Trigeminal ganglion
INFRAORBITAL NERVE through infraorbital foramen
Maxillary (V_2) nerve
SUPERIOR ALVEOLAR NERVES: anterior, middle, and posterior (cut)
PTERYGOPALATINE NERVE AND GANGLION
Pharyngeal nerve
Nasopalatine nerve
LESSER PALATINE NERVES (lesser and middle)
GREATER PALATINE NERVE
L Wilson-Pauwels

FIGURE V–6 Palatine nerves (inferior view)

maxilla and travels posteriorly through the infraorbital canal where it is joined by anterior branches of the superior alveolar nerve. This combined trunk emerges on the floor of the orbit and joins the maxillary nerve. The maxillary nerve continues posteriorly and is joined by the middle and posterior branches of the superior alveolar nerves and by the palatine nerves. The combined trunk, the maxillary division, enters the cranium through the foramen rotundum.

The *superior alveolar nerves* (anterior, middle, and posterior) carry sensory input, mainly pain, from the upper teeth.

The *palatine nerves* (see Fig. V–6) (greater and lesser) originate in the hard and soft palates, respectively, and ascend toward the maxillary nerve through the pterygopalatine canal. En route, the palatine nerves are joined by a pharyngeal branch from the nasopharynx and by nasal branches from the posterior nasal cavity, including one particularly long branch, the *nasopalatine nerve*.

Small meningeal branches from the dura of the anterior and middle cranial fossae join the maxillary division as it enters the trigeminal ganglion (see Fig. V–5).

Mandibular Division (V_3)

The sensory component of V_3 is formed by the buccal, lingual, inferior alveolar, and auriculotemporal nerves (Fig. V–7).

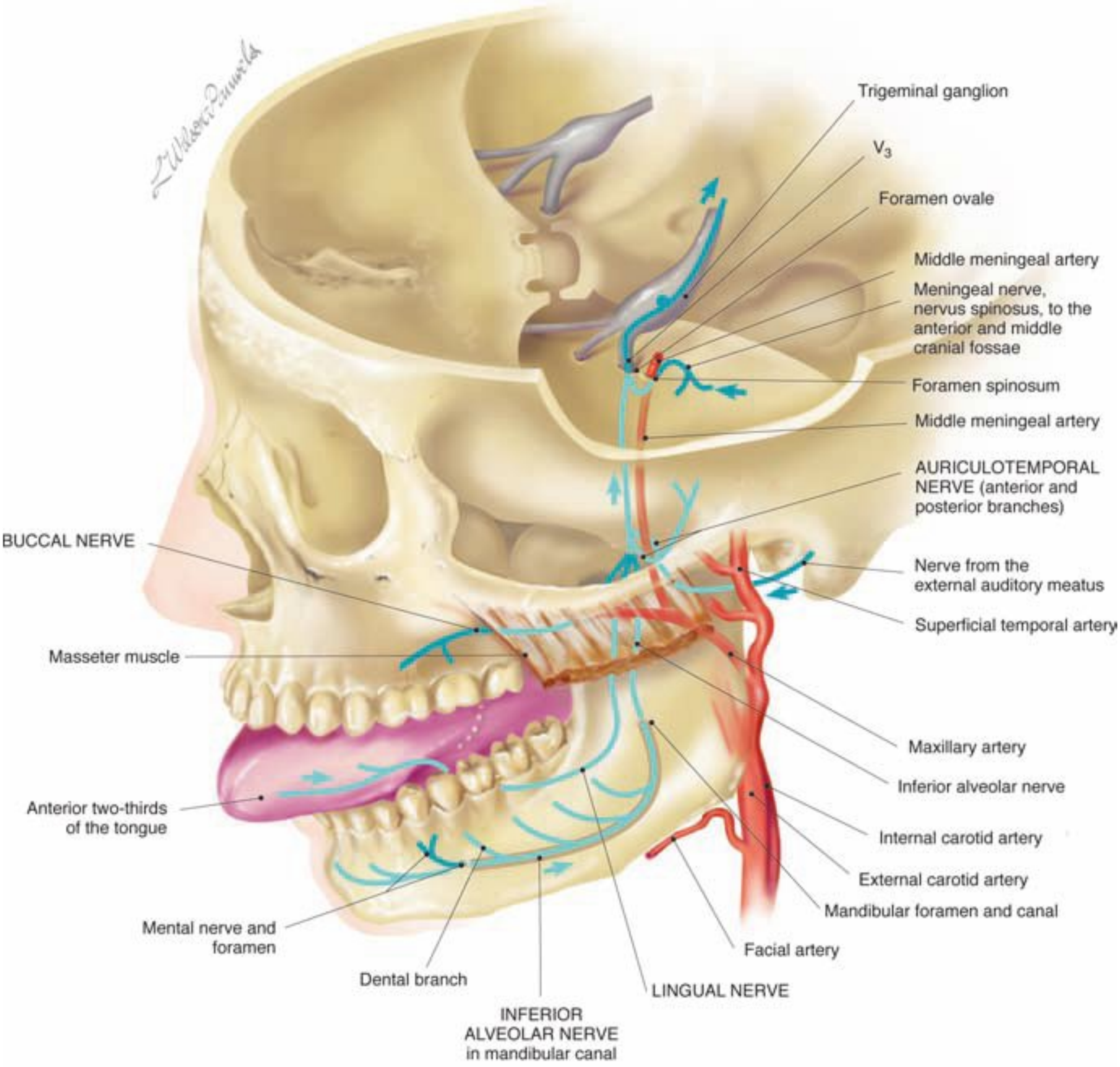
Trigeminal ganglion
V_3
Foramen ovale
Middle meningeal artery
Meningeal nerve, nervus spinosus, to the anterior and middle cranial fossae
Foramen spinosum
Middle meningeal artery
AURICULOTEMPORAL NERVE (anterior and posterior branches)
BUCCAL NERVE
Nerve from the external auditory meatus
Superficial temporal artery
Masseter muscle
Maxillary artery
Inferior alveolar nerve
Anterior two-thirds of the tongue
Internal carotid artery
External carotid artery
Mandibular foramen and canal
Mental nerve and foramen
Facial artery
Dental branch
LINGUAL NERVE
INFERIOR ALVEOLAR NERVE in mandibular canal

FIGURE V–7 General sensory component of the trigeminal nerve–mandibular (V_3) division.

The *buccal nerve* (not to be confused with the nerve to buccinator, a motor branch of CN VII) carries sensory information from the buccal (cheek) region including the mucous membrane of the mouth and gums. The *buccal nerve* courses posteriorly in the cheek deep to the masseter muscle and pierces the lateral pterygoid muscle to join the main trunk of the mandibular nerve.

Sensory axons from the anterior two-thirds of the tongue converge to form the *lingual nerve*, which runs posteriorly along the side of the tongue. At the back of the tongue, the lingual nerve curves upward to join the main trunk of the mandibular nerve deep to the lateral pterygoid muscle.

Sensory nerves from the chin and lower lip converge to form the mental nerve, which enters the mandible through the mental foramen to run in the mandibular canal. Within the canal, dental branches from the lower teeth join with the mental nerve to form the *inferior alveolar nerve*. This nerve continues posteriorly and exits from the mandibular canal through the mandibular foramen to join the main trunk of the mandibular division along with the lingual nerve.

The *auriculotemporal nerve*, which runs with the superficial temporal artery, carries sensation from the side of the head and scalp. Two main branches, the anterior and posterior auriculotemporal nerves and their tributaries, converge into a single trunk just anterior to the ear. Here they are joined by twigs from the external auditory meatus, the external surface of the tympanic membrane, and the temporomandibular joint. The nerve courses deep to the lateral pterygoid muscle and the neck of the mandible, splits to encircle the middle meningeal artery, and then joins the main trunk of the mandibular nerve. The entire mandibular division enters the cranium through the foramen ovale.

All three divisions of the trigeminal nerve, ophthalmic, maxillary, and mandibular, join together at the trigeminal ganglion where most of the sensory nerve cell bodies reside. Central processes of these neurons constitute the sensory root of the trigeminal nerve, which enters the pons at its midlateral point. The axons terminate by synapsing with second-order sensory neurons in the appropriate region of the trigeminal nucleus.

The Trigeminal Nucleus

The trigeminal sensory nucleus is the largest of the CN nuclei. It extends from the midbrain caudally into the spinal cord as far as the second cervical segment where it becomes continuous with the dorsal horn of the spinal cord (Fig. V–8). Within the medulla, it creates a lateral elevation—the tuberculum cinereum. It has three subnuclei: the mesencephalic trigeminal nucleus, the pontine trigeminal (chief sensory) nucleus, and the spinal trigeminal nucleus (Figs. V–9 and V–10; see also Fig. V–8).

The mesencephalic trigeminal nucleus is a unique structure. The neurons of this nucleus are first-order sensory neurons; however, they are located within the central nervous system (CNS), whereas all other first-order sensory neurons are located in ganglia outside of the CNS. Some authors consider the mesencephalic trigeminal neurons to be a displaced part of the trigeminal ganglion. The significance of this arrangement is not well understood. These neurons form a very thin column extending into the rostral midbrain. Their peripheral processes form the tract of the mesencephalic trigeminal nucleus, just lateral to the cell column. They carry signals from muscle spindles in the muscles of mastication and from mechanoreceptors

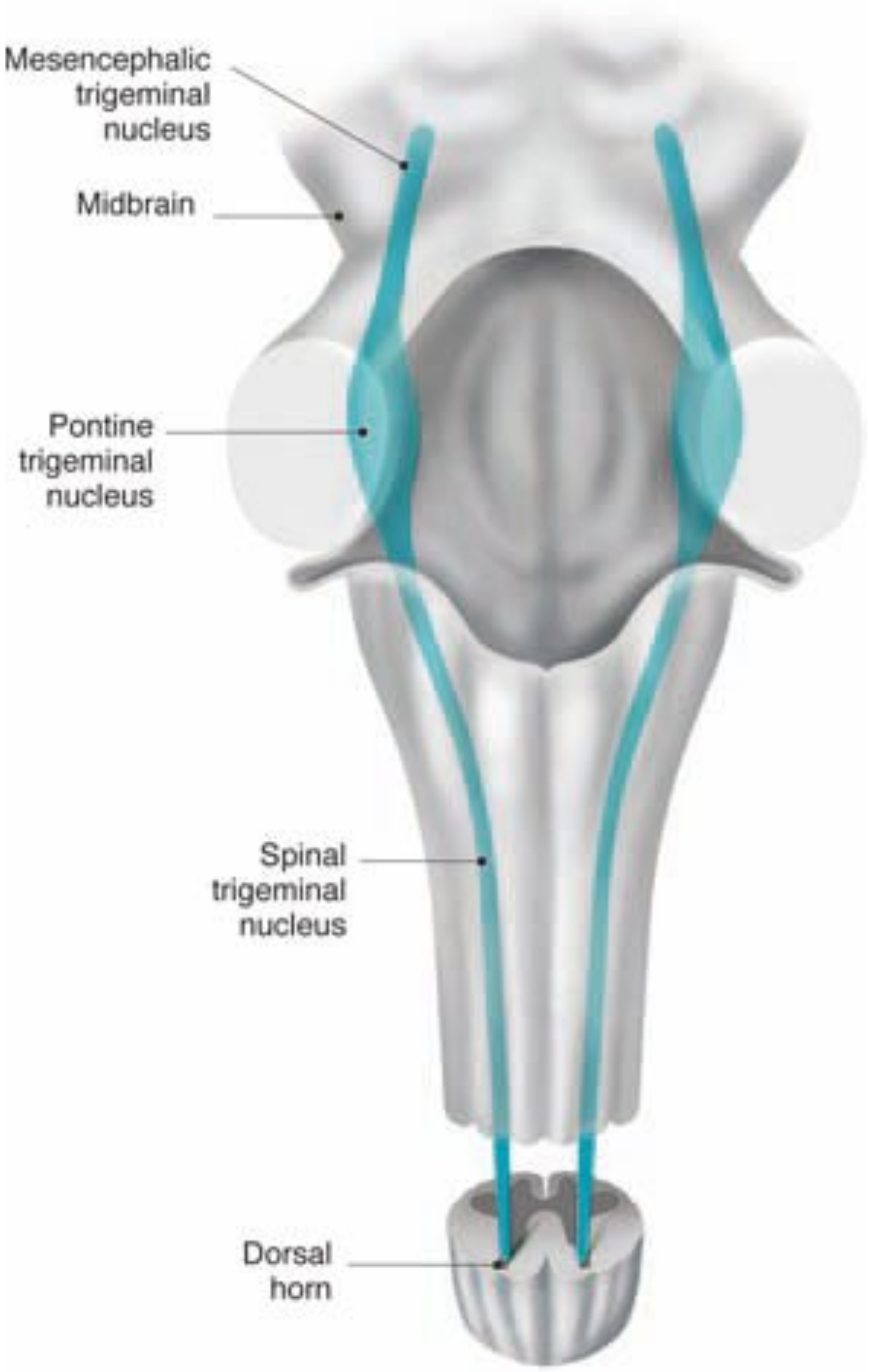
Mesencephalic trigeminal nucleus
Midbrain
Pontine trigeminal nucleus
Spinal trigeminal nucleus
Dorsal horn

FIGURE V–8 Trigeminal sensory nucleus (dorsal view of the brain stem).

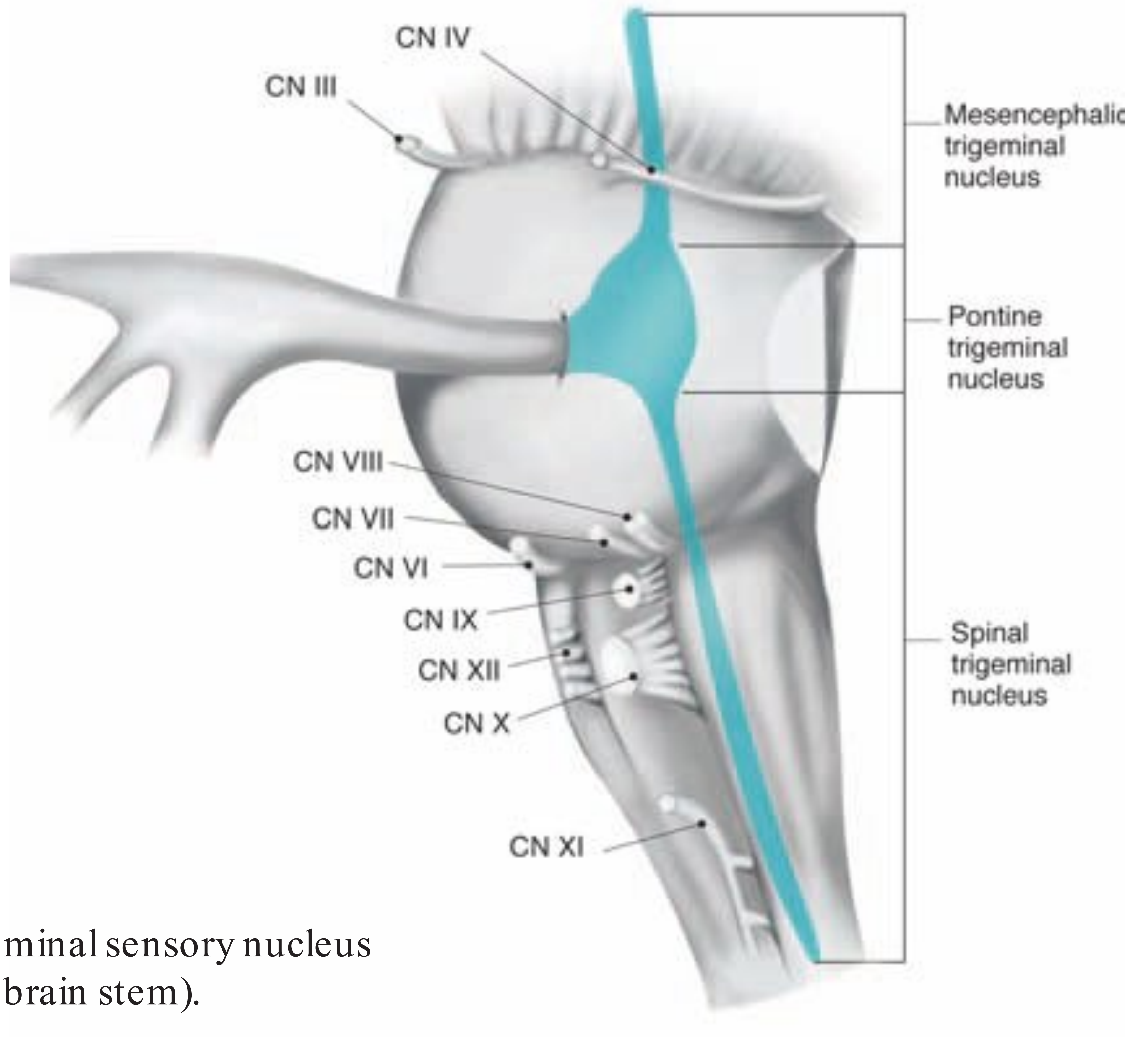
CN IV
CN III
Mesencephalic trigeminal nucleus
Pontine trigeminal nucleus
CN VIII
CN VII
CN VI
CN IX
CN XII
CN X
Spinal trigeminal nucleus
CN XI

FIGURE V–9 Trigeminal sensory nucleus (lateral view of the brain stem).

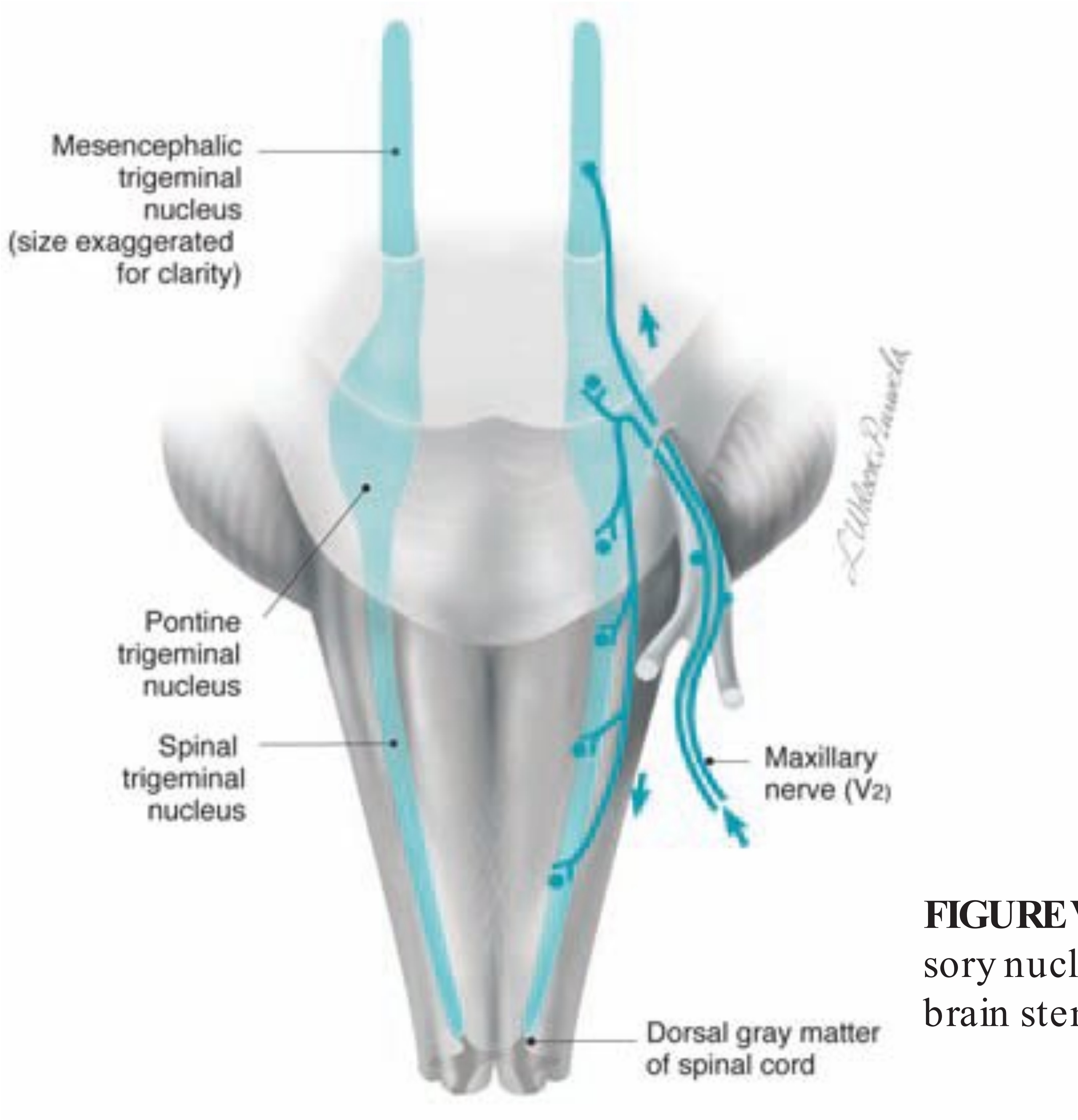
Mesencephalic trigeminal nucleus (size exaggerated for clarity)
Pontine trigeminal nucleus
Spinal trigeminal nucleus
Maxillary nerve (V2)
Dorsal gray matter of spinal cord

FIGURE V–10 Trigeminal sensory nucleus (ventral view of the brain stem).

in gums, teeth, and hard palate. The central processes project mainly to the motor nucleus of CN V (masticator nucleus) to provide for reflex control of the bite (Fig. V–11). These neurons form the afferent limb of the mandibular (jaw jerk) reflex described later in "Clinical Testing."

The pontine trigeminal nucleus (also called the "chief sensory nucleus") is a large group of secondary sensory neurons located in the pons near the point of entry of the nerve. It is concerned primarily with discriminative touch sensation from the face.

The pontine trigeminal nucleus is composed of two parts (Fig. V–12 inset).

- The dorsomedial part is concerned primarily with information from the oral cavity. It is the origin of the ipsilateral dorsal (posterior) trigeminothalamic tract described later in "Central Pathways."
- The ventrolateral part receives signals from all of the trigeminal divisions and is somatotopically organized. The mandibular division is represented most dorsally, the ophthalmic division is represented most ventrally, and the maxillary division is represented in between. These neurons give rise to the crossed projection to the contralateral thalamus, described later.

The spinal trigeminal nucleus is a long, substantial column of cells extending from the pontine trigeminal nucleus caudally into the spinal cord as far as C2, where it

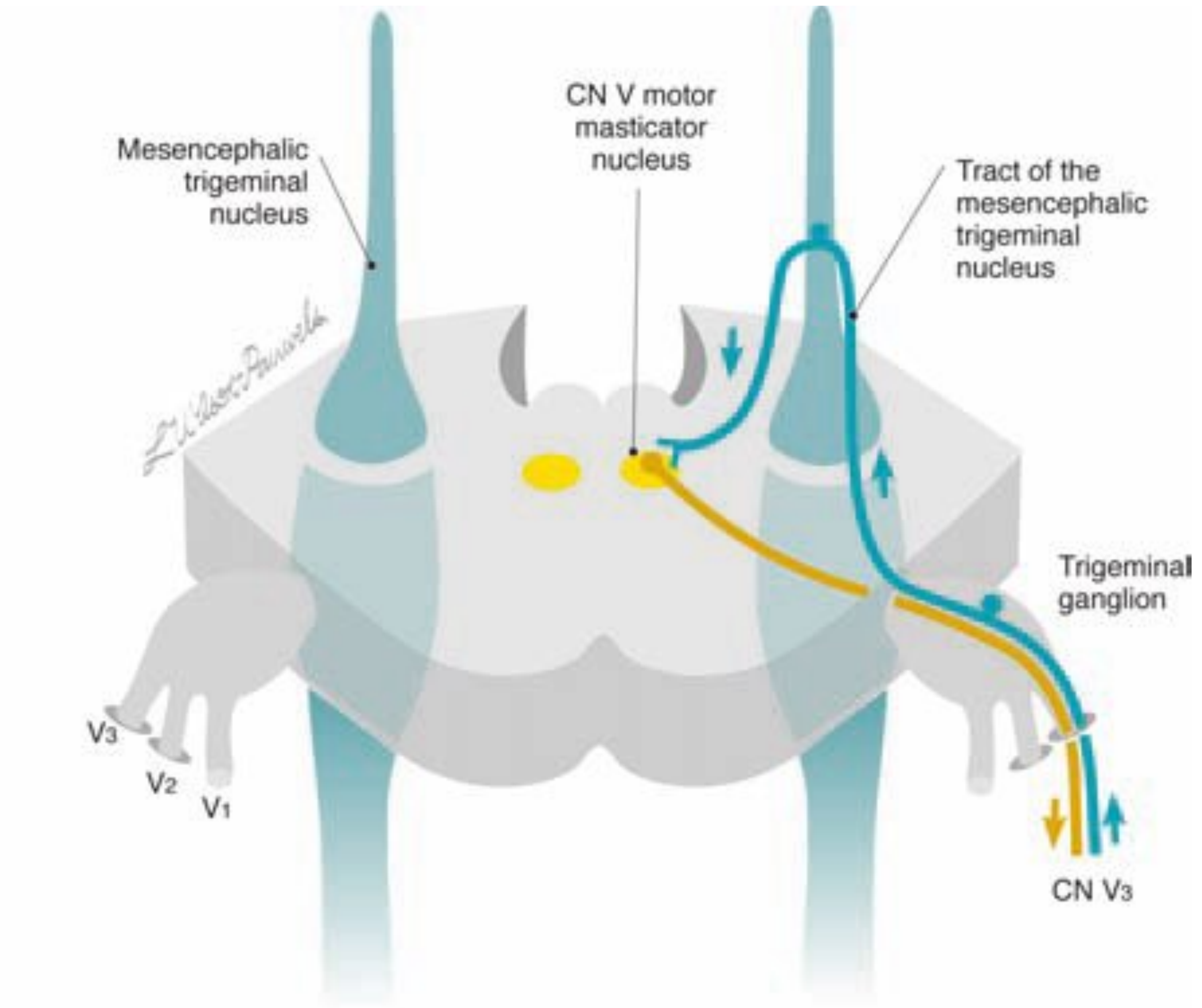

FIGURE V–11 Mandibular jaw reflex (sensory input V_3, motor output to muscles of mastication V_3).

merges with the dorsal gray matter of the spinal cord (see Figs. V–8 and V–10). The central processes of the spinal trigeminal ganglion neurons form a sizeable tract on the lateral aspect of the nucleus, called the "tract of the spinal trigeminal nucleus."

This subnucleus, especially its caudal portion, is concerned primarily with relaying pain and temperature signals, although some tactile information is projected here as well as to the pontine trigeminal nucleus. Axons of spinal trigeminal nucleus neurons project to the contralateral sensory cortex via the thalamus. Like the pontine sensory nucleus, the caudal part of the spinal trigeminal nucleus and its tract are also organized somatotopically. The ophthalmic division is represented most ventrally in the tract and nucleus, the mandibular division is represented most dorsally, and the maxillary division is sandwiched in between.

CENTRAL PATHWAYS

Two major sensory pathways carry sensations from the face and sinuses to the cerebrum: the discriminative touch pathway (see Fig. V–12) and the pain and temperature pathway (Fig. V–13).

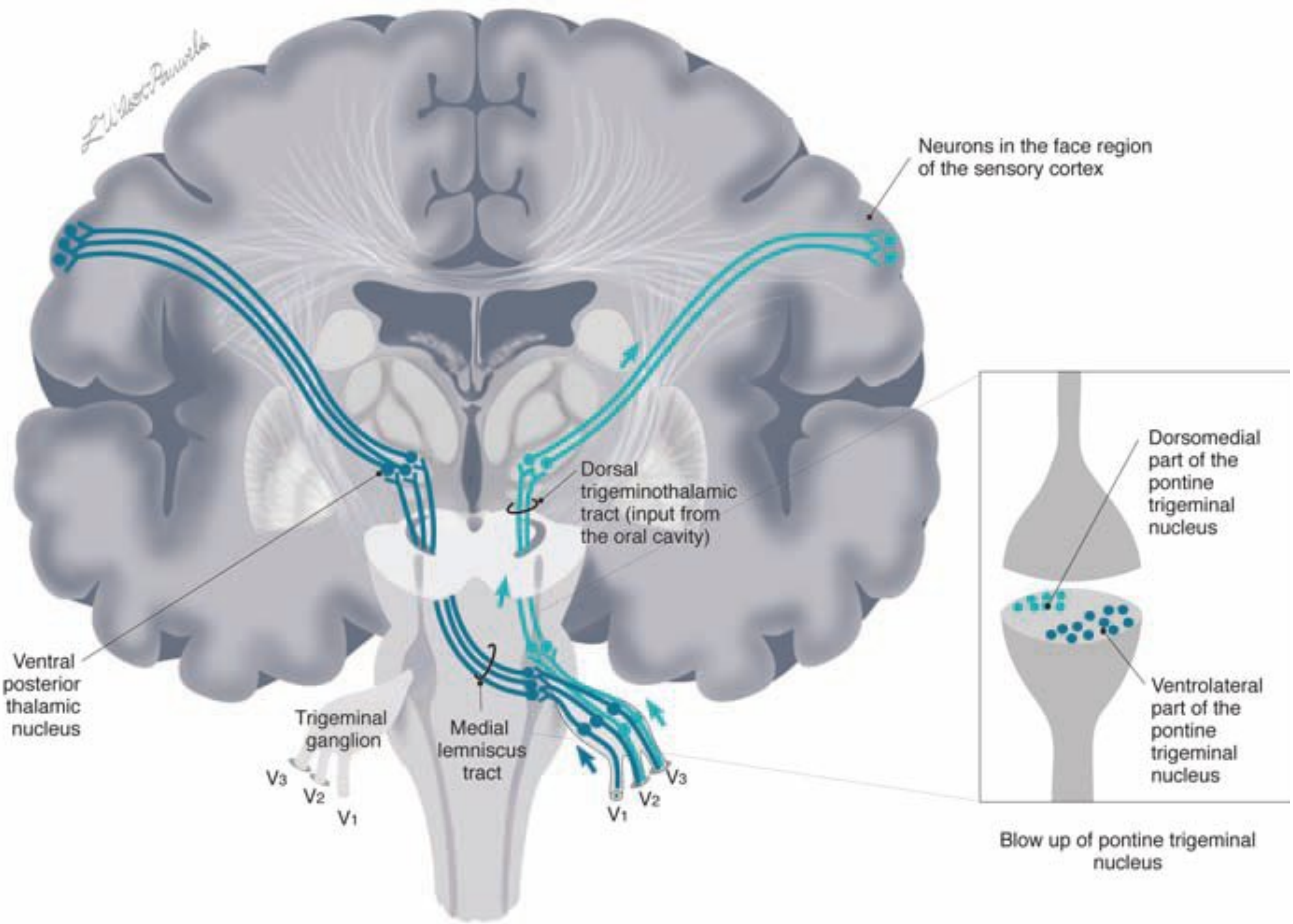
L. Wilson-Pauwels
Neurons in the face region of the sensory cortex
Dorsomedial part of the pontine trigeminal nucleus
Dorsal trigeminothalamic tract (input from the oral cavity)
Ventral posterior thalamic nucleus
Trigeminal ganglion
Medial lemniscus tract
V3
V2
V1
V1
V2
V3
Ventrolateral part of the pontine trigeminal nucleus
Blow up of pontine trigeminal nucleus

FIGURE V–12 The discriminative touch pathway from the head. Sensory signals from the oral cavity via V_2 and V_3 project from the dorsomedial part of the pontine trigeminal nucleus via the dorsal trigeminothalamic tract to the ipsilateral ventral posterior thalamus and the sensory cortex. Sensory signals from the remaining areas of the head (V_1, V_2, V_3) project from the ventrolateral part of the pontine trigeminal nucleus via the medial lemniscus tract to the contralateral ventral posterior thalamus and the sensory cortex.

Discriminative Touch Pathway

The discriminative touch pathway carries the sensory modalities of two-point discrimination: vibration sensation and proprioception. In terms of evolution, this pathway is a recent addition to the nervous system and is highly developed in primates.

Like most sensory pathways, the discriminative touch pathway includes three principal neurons (see Fig. V–12).

1. The *first-order sensory neurons* carry information from a specific region of the face or meninges to the pontine trigeminal nucleus.

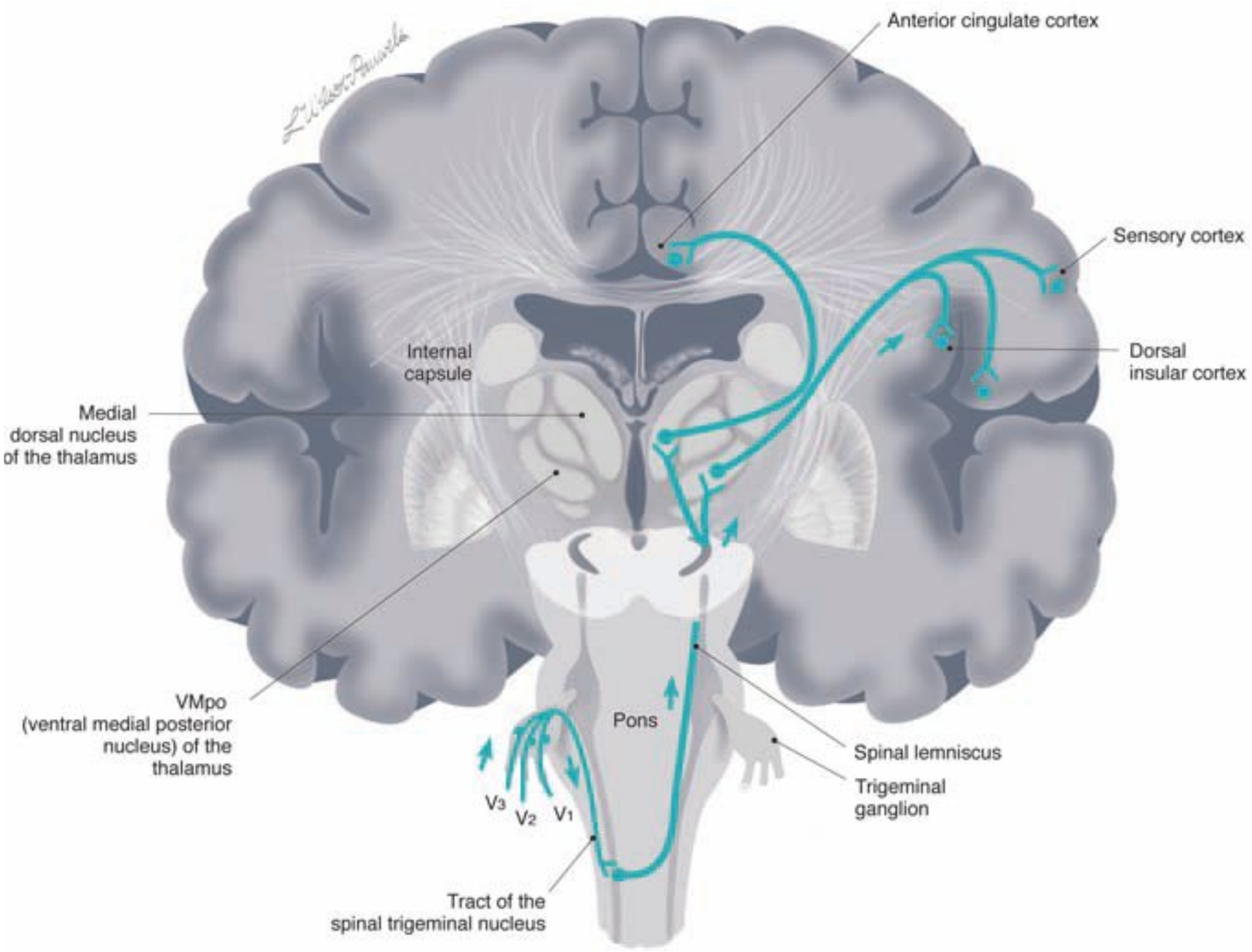
Anterior cingulate cortex
Sensory cortex
Dorsal insular cortex
Internal capsule
Medial dorsal nucleus of the thalamus
VMpo (ventral medial posterior nucleus) of the thalamus
Pons
Spinal lemniscus
Trigeminal ganglion
V3
V2
V1
Tract of the spinal trigeminal nucleus
L Wilson-Pauwels

FIGURE V–13 Pain and temperature pathway from the head (pathways after Dostrovsky and Craig 2006).

2. The pontine trigeminal nucleus is composed of cell bodies of *second-order sensory neurons*. These neurons give rise to two ascending tracts:

   The dorsomedial part of the nucleus receives signals from the oral cavity. Axons of these neurons form the dorsal (posterior) trigeminothalamic tract, which terminates in the medial aspect of the ipsilateral ventral posterior thalamic nucleus. Interestingly, this is adjacent to the thalamic area where taste signals (CNs VII and IX) from the ipsilateral side of the mouth terminate.

   The ventrolateral part of the nucleus receives signals from all divisions of the trigeminal nerve. Axons leave this nucleus centrally and cross the midline to join the medial lemniscus en route to the thalamus where they terminate within the medial aspect of the ventral posterior thalamic nucleus. Some authors distinguish these fibers as the ventral (anterior) trigeminothalamic tract.

3. Cell bodies of the *third-order sensory neurons* reside in the medial aspect of the ventral posterior nucleus of the thalamus. Their axons leave the thalamus and travel through the posterior limb of the internal capsule and corona radiata to terminate in the face region of the primary sensory cortex where sensory signals are consciously perceived.

Pain and Temperature Pathway

Pain and temperature sensation is carried by a much more primitive, widespread pathway than the discriminative touch pathway. In addition to the objective localization of pain sensation, the central pathways provide for activation of the limbic system and the activation of the fight-or-flight response that may be an appropriate response to pain. Pain is usually distinguished as "fast" and "slow." *Fast pain* is carried on small myelinated axons to the CNS. Central projections in this pathway provide for localizing the pain and evaluating its intensity. *Slow pain* is carried to the CNS by unmyelinated, slower-conducting axons.* Central projections in this pathway provide for the suffering, distressing aspects of pain.

The trigeminal pain and temperature pathway also includes three principal neurons (see Fig. V–13).

1. The *primary* or *first-order neurons*, both myelinated and unmyelinated, carry impulses from the periphery to the CNS. Their cell bodies are located in the trigeminal ganglion, and their central processes enter the pons along with the central processes of the discriminative touch neurons. Once they have entered the brain stem, they turn caudally and descend within the brain stem, forming the tract of the spinal trigeminal nucleus, before terminating within the appropriate parts of the nucleus.
2. Cell bodies of the *second-order sensory neuron* form the spinal trigeminal nucleus. Their axons cross the midline and join the spinal lemniscus en route to the thalamus. Some authors consider these axons to be part of the ventral (anterior) spinal trigeminal lemniscus. En route to the thalamus, these axons send collateral branches to the reticular formation in the brain stem, to activate arousal and visceral responses to pain, and to the periaqueductal gray area, to activate descending, pain-inhibiting pathways.

   Pain and temperature axons terminate in two areas of the thalamus (see Fig. V–13).

   - A recently distinguished nucleus in the posterolateral thalamus, the ventral medial posterior nucleus (VMpo). This nucleus is well developed only in humans, and it is the primary target for spinal lemniscus axons.
   - The medial dorsal nucleus of the thalamus.

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*Perception of these two sets of signals can be separated by 1 or 2 seconds if they originate in a distant part of the body, such as the foot.

3. The thalamic nuclei involved in relaying sensory information to the cortex are composed of *third-order (thalamic) neurons*. Neurons of the VMpo project to the sensory cortex and to the posterior dorsal insular cortex where the sensory signals are consciously perceived in terms of their location and intensity. Neurons in the medial dorsal nucleus of the thalamus project to the anterior cingulate cortex where the distressing, emotional aspects of pain are mediated.

Light or simple touch, is poorly localized and is probably carried by both the discriminative touch, and the pain and temperature pathways.

BRANCHIAL MOTOR (EFFERENT) COMPONENT

Central Motor Nuclei: Innervation of the Masticator Nucleus

The trigeminal motor (masticator) nuclei are located in the tegmentum of the pons, medial to the pontine trigeminal nuclei (Fig. V–14). They innervate the muscles of mastication (i.e., masseters, temporales, medial and lateral pterygoid muscles), plus tensores tympani, tensores veli palatini, mylohyoid muscles, and anterior bellies of the digastric muscles.

In humans, jaw movements have two functions: a primitive function in masticating food and a more modern function in articulating speech sounds. These motor neurons, therefore, receive signals from two distinct sources:

- Because chewing is primarily a reflex activity in response to sensory signals from the mouth, the masticator nucleus receives substantial inputs from the sensory nuclei of the trigeminal nerve, both directly and via the pontine reticular formation.
- In addition, it receives signals from both cerebral hemispheres via the corticobulbar tract that mediate the jaw movements involved in speech and voluntary chewing.

Specialist Comment

Input from the vestibulocochlear nerve activates the part of the trigeminal motor nucleus that innervates the tensor tympani, so that the tension of the tympanic membrane (ear drum) can be adjusted for sound intensity (see Chapter VIII, Fig. VIII–12).

Peripheral Nerves

The motor nerves to the muscles of mastication travel with the mandibular division of the trigeminal nerve (Fig. V–15; see also Fig. V–14). The masticator nucleus receives bilateral innervation from the cortices. Axons from the masticator nucleus (LMNs) course laterally through the pons to exit as the motor root on the medial

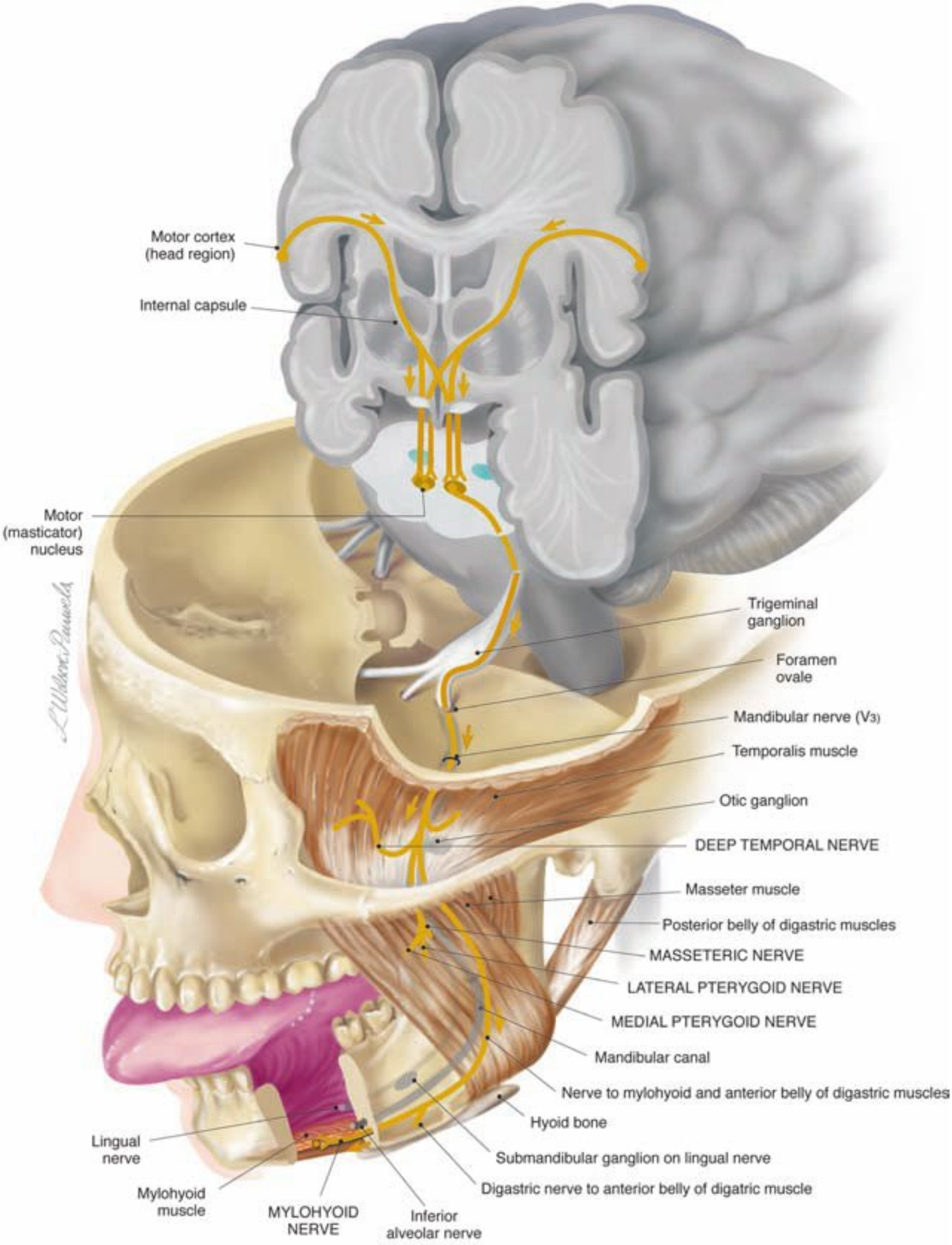

FIGURE V–14 The branchial motor component of the trigeminal nerve.

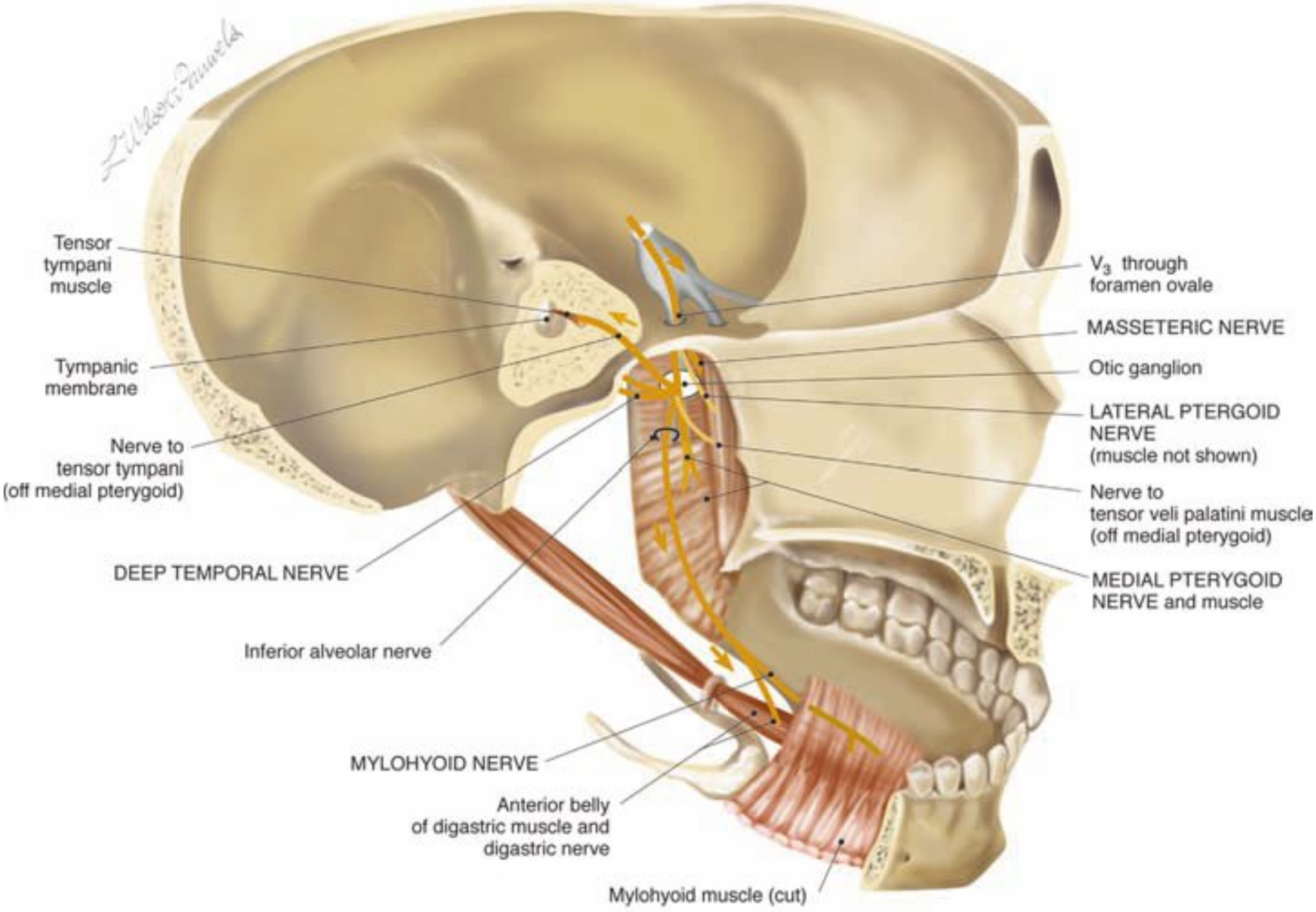
L. Wilson-Pauwels
Tensor tympani muscle
Tympanic membrane
Nerve to tensor tympani (off medial pterygoid)
DEEP TEMPORAL NERVE
Inferior alveolar nerve
MYLOHYOID NERVE
Anterior belly of digastric muscle and digastric nerve
Mylohyoid muscle (cut)
V_3 through foramen ovale
MASSETERIC NERVE
Otic ganglion
LATERAL PTERGOID NERVE (muscle not shown)
Nerve to tensor veli palatini muscle (off medial pterygoid)
MEDIAL PTERYGOID NERVE and muscle

FIGURE V–15 Deep branches of the trigeminal motor division viewed from the medial aspect of the left mandible.

aspect of the sensory trigeminal root (V_3). The motor axons course deep to the trigeminal ganglion in the middle cranial fossa and leave the cranium through the foramen ovale (see Fig. V–14). Just outside the foramen ovale, they divide into five main branches: the medial and lateral pterygoid nerves, the masseteric nerve, the deep temporal nerves, and the mylohyoid nerve.

The *medial pterygoid nerve* gives off two small branches to the tensor veli palatini and tensor tympani and then enters the deep surface of the medial pterygoid muscle, which it supplies (see Fig. V–15).

The *lateral pterygoid nerve* runs briefly with the buccal nerve and enters the deep surface of the lateral pterygoid muscle.

The *masseteric nerve* passes laterally above the lateral pterygoid muscle through the mandibular notch to supply the masseter (see Fig. V–14).

Two to three *deep temporal nerves* branch from the mandibular nerve, turn upward, and pass superior to the lateral pterygoid muscle to enter the deep surface of the temporalis muscle (see Fig. V–14).

The *mylohyoid nerve* travels with the inferior alveolar nerve, branching from it just before the latter enters the mandibular canal. The mylohyoid nerve continues anteriorly and inferiorly in a groove on the deep surface of the ramus of the mandible to reach the inferior surface of the mylohyoid muscle where it divides to supply the anterior belly of the digastric muscle and the mylohyoid muscle (see Fig. V–15).

CASE HISTORY GUIDING QUESTIONS

1. What is trigeminal neuralgia?
2. What was Mary's family doctor looking for on the MRI scans?
3. Why are anticonvulsants effective in the treatment of trigeminal neuralgia?
4. Where along the trigeminal sensory and motor pathways can other pathologies occur?
5. What is the blink reflex, and why is it important?

1. What is trigeminal neuralgia?

Trigeminal neuralgia, also known as "tic douloureux" (painful spasm), is a condition in which excruciating paroxysms of pain occur in one or more divisions of the trigeminal nerve. The pain is typically lancinating in character and lasts a few seconds. The mandibular or maxillary divisions of the trigeminal nerve are most often involved, whereas the ophthalmic division is rarely involved. It is one of the most severe pains known in clinical medicine. The pain can occur spontaneously or can be elicited by eating, talking, shaving, brushing the teeth, or touching a trigger zone. Trigeminal neuralgia patients often avoid these activities. To reduce sensory input from the mouth, many stop eating and may lose weight. They also protect their faces to avoid inadvertent touching. When the condition is severe, pain can occur many times an hour. Between attacks there is no pain, although patients live in dread of the next attack. Trigeminal neuralgia has an estimated incidence of 15 to 100 per 100,000 population. Women are twice as likely as men to develop this disease.

The precise mechanism of pain production is unknown, but in some cases, it is thought to be due to local irritation of the sensory fibers in the trigeminal nerve, which gives rise to spontaneous, ectopic impulses. A short high-frequency barrage of impulses is then projected to the cortex, giving rise to the perception of severe pain.

2. What was Mary's family doctor looking for on the MRI scans?

Mary's doctor suspected that something was pressing on the root of her left trigeminal nerve causing the ectopic impulses that gave rise to her acutely painful

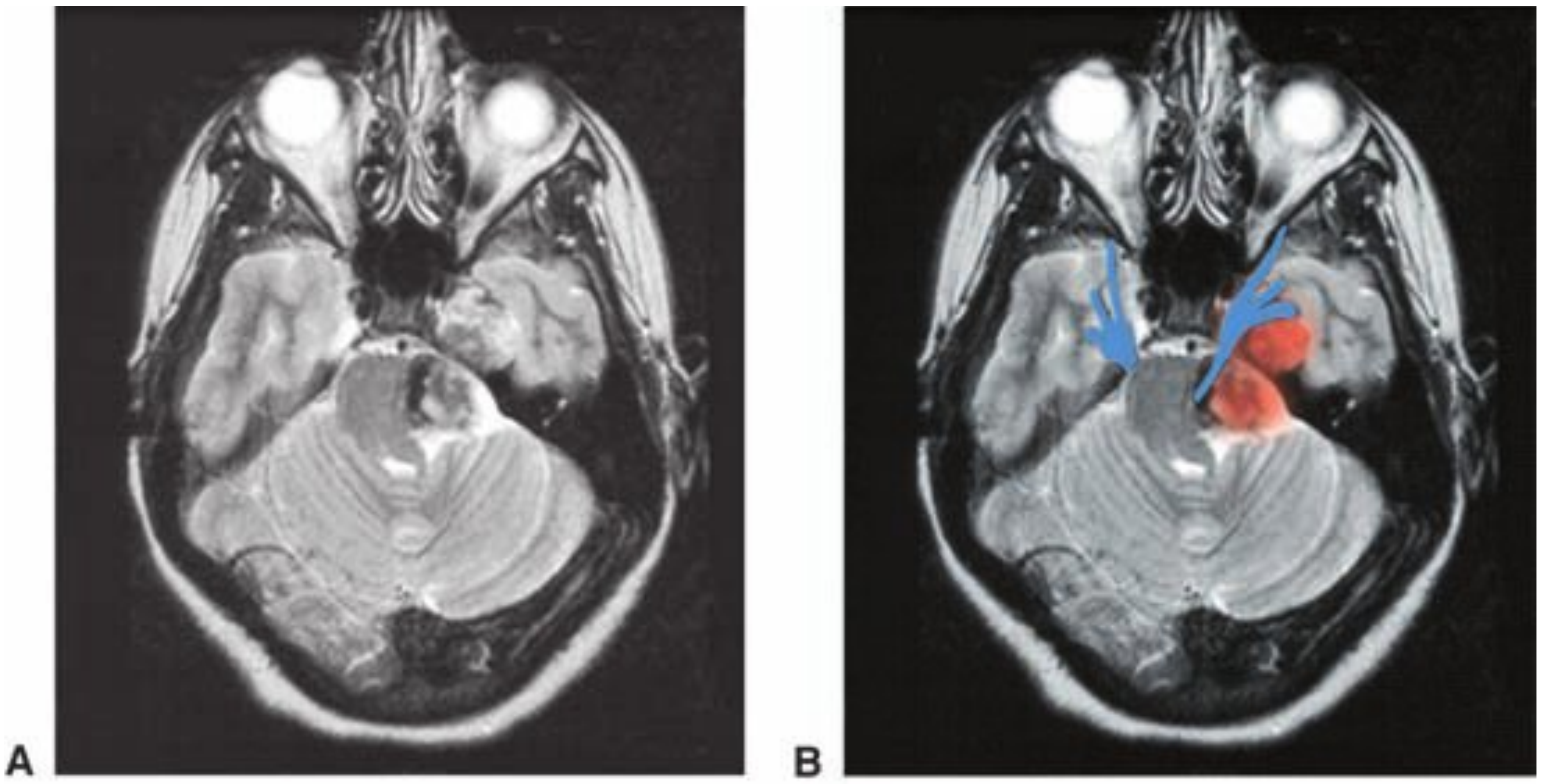

FIGURE V–16 Mary's MRI showed a meningioma growing on her left petrous temporal ridge. The tumor is pressing on her pons at the entry of the trigeminal nerve. **A**, MRI scan. **B**, The tumor has been artifically coloured (red) and representations of the positions of the trigeminal nerves have been ghosted in to show that the left trigeminal nerve (blue) would be compressed by the meningioma. (Image courtesy of Dr. David Mikulis.)

sensations. The MRI scan (Fig. V–16) showed a small meningioma in the middle cranial fossa adjacent to her trigeminal nerve root, which was the likely cause of her pain. When tumors are present, trigeminal neuralgia may be associated with other neurologic findings because other anatomic structures in the brain stem are in close proximity and are easily compressed. In other cases, the trigeminal nerve can be irritated by the pulsations of a tortuous superior cerebellar artery.

3. Why are anticonvulsants effective in the treatment of trigeminal neuralgia?

Anticonvulsants act to decrease the firing rate of neurons. Carbamazepine is a sodium channel blocker that exerts its effect by decreasing the neuron's ability to produce a rapid train of impulses. Mary does not have epilepsy but, like epileptic patients, she does have an abnormal neuronal excitability. Mary's trigeminal neurons are hyperexcited because of the irritation caused by the meningeal tumor.

4. Where along the trigeminal sensory and motor pathways can other pathologies occur?

Pathology in the trigeminal system can occur anywhere along the course of the peripheral nerves, within the middle cranial fossa, or within the central nervous system.

- *Along the Course of the Peripheral Nerves*

  Branches of the trigeminal nerve can be damaged by injury to the head. The sensory and/or motor loss suffered depends on which branches are damaged. Herpes zoster (a virus infection commonly referred to as "shingles") can invade the trigeminal ganglion where it can lie dormant. It can become active and travel along the peripheral processes, which leads to irritation during the outbreak and pain following resolution.

- *Within the Middle Cranial Fossa*

  As in Mary's case, the peripheral nerve can be damaged from compression by a meningioma as the nerve passes through the middle cranial fossa or by a schwannoma of the nerve itself. Schwannomas of the acoustic nerve, if sufficiently large, can also affect the trigeminal nerve, causing pain and eventual loss of function. Because of their anatomic location at the junction of the cerebellum and the pons, schwannomas give rise to a collection of symptoms (including trigeminal dysfunction) known as the "cerebellopontine angle syndrome" (Fig. V–17) (see also Chapter VIII).

- *Within the Central Nervous System*

  Tumors, demylinating disease (multiple sclerosis), and vascular problems can affect trigeminal pathways within the CNS.

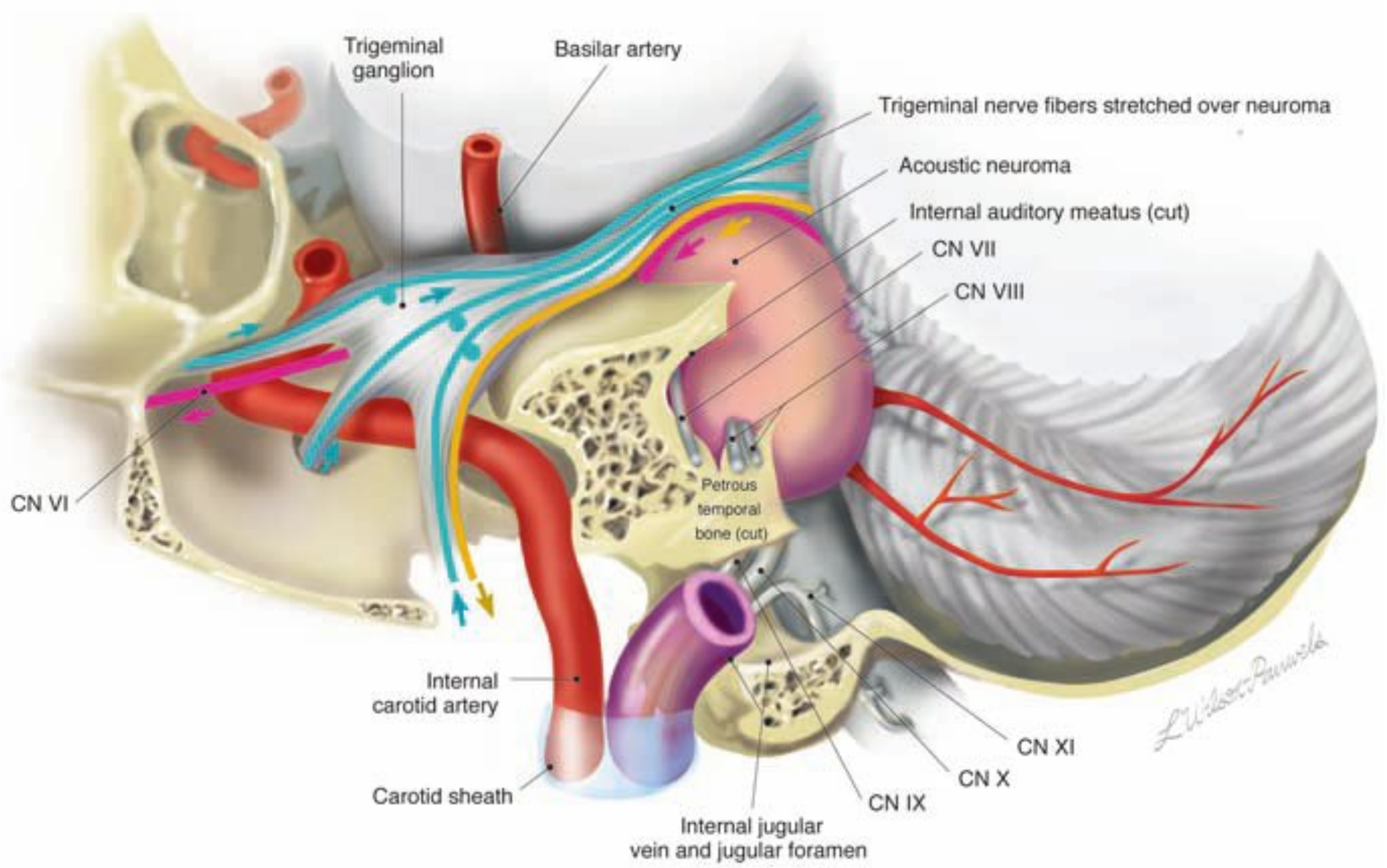
Trigeminal ganglion
Basilar artery
Trigeminal nerve fibers stretched over neuroma
Acoustic neuroma
Internal auditory meatus (cut)
CN VII
CN VIII
CN VI
Petrous temporal bone (cut)
Internal carotid artery
Carotid sheath
Internal jugular vein and jugular foramen
CN IX
CN X
CN XI

FIGURE V–17 Schwannoma: an enlarged tumor in the cerebellopontine angle compressing the root of the trigeminal nerve (sagittal section through the jugular foramen).

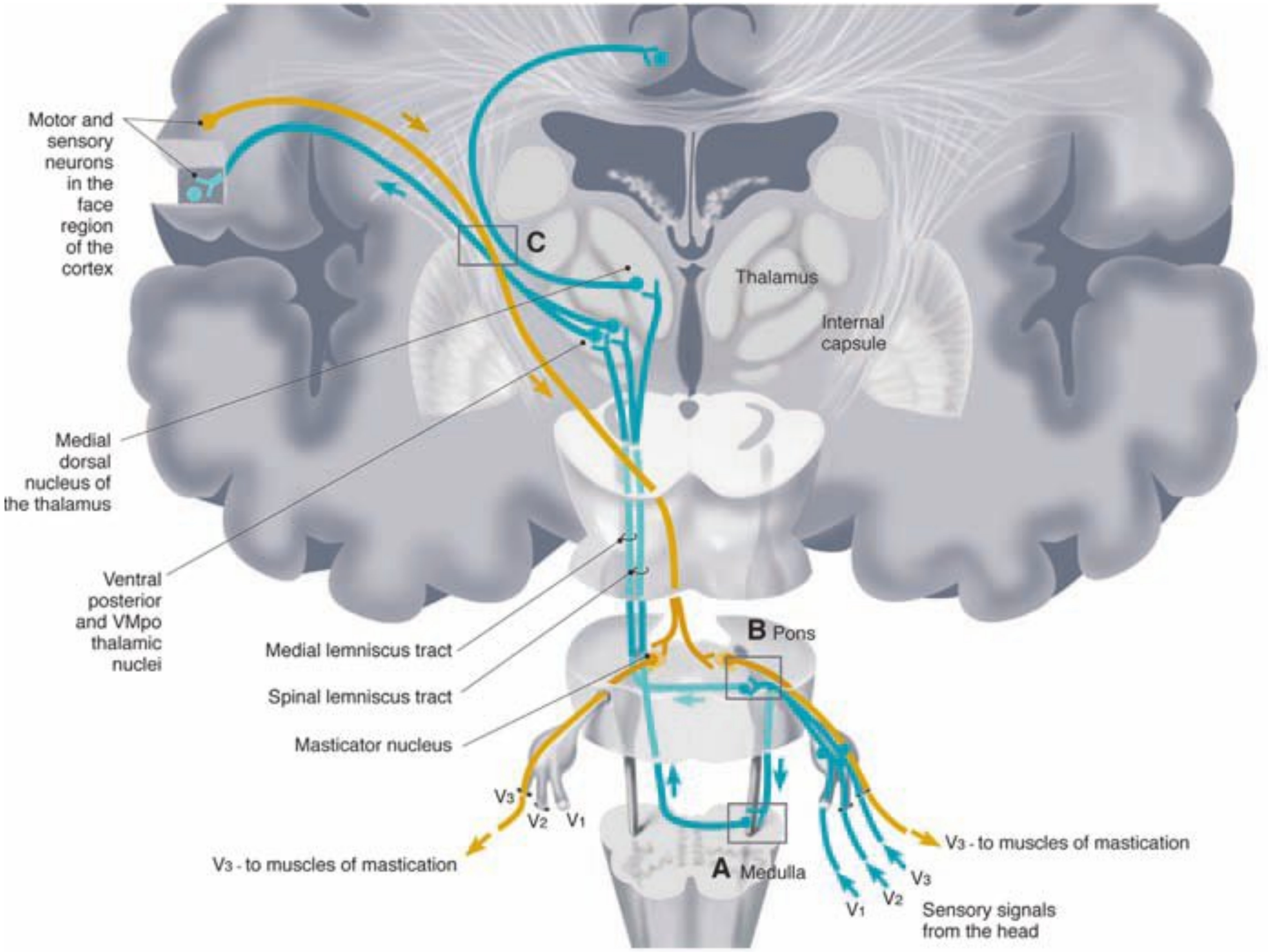
Motor and sensory neurons in the face region of the cortex
C
Thalamus
Internal capsule
Medial dorsal nucleus of the thalamus
Ventral posterior and VMpo thalamic nuclei
Medial lemniscus tract
Spinal lemniscus tract
Masticator nucleus
B Pons
V3
V2
V1
V3 - to muscles of mastication
A Medulla
V3 - to muscles of mastication
V3
V2
V1
Sensory signals from the head

FIGURE V–18 Damage to the trigeminal pathways. **A**, Within the medulla; **B**, within the pons; and **C**, above the brain stem (contralateral tract). See the text for a description of the functional loss that would result from lesions **A**, **B** and **C**.

- Within the medulla, damage to the trigeminal system gives rise primarily to loss of pain and temperature sensation on the ipsilateral (same) side of the face (Fig. V–18A).
- Within the pons, ipsilateral discriminative touch is primarily affected. Innervation of the ipsilateral muscles of mastication may also be affected, giving rise to a lower motor neuron lesion (see Fig. V–18B).
- At levels of the neuraxis above the brain stem (see Fig. V–18C), all sensory modalities on the contralateral (opposite) side of the head are affected. Motor function is not affected because the motor (masticator) nucleus is driven by sensory reflex input from sensory neurons at the same level (see blink reflex, Fig. V–19) and because it receives bilateral innervation from the cerebral hemispheres.

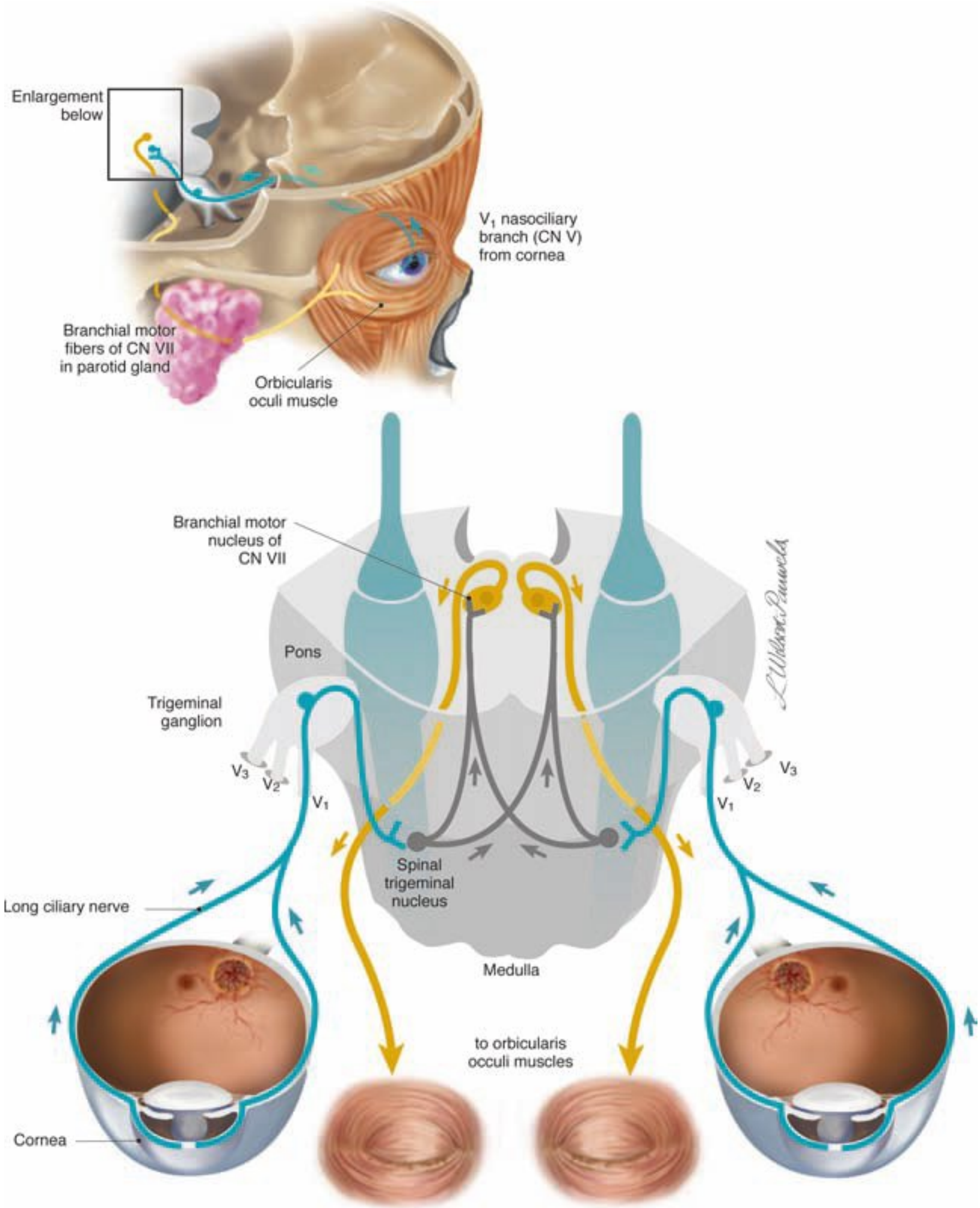
Enlargement below
V_1 nasociliary branch (CN V) from cornea
Branchial motor fibers of CN VII in parotid gland
Orbicularis oculi muscle
Branchial motor nucleus of CN VII
Pons
Trigeminal ganglion
V_3
V_2
V_1
V_3
V_2
V_1
Spinal trigeminal nucleus
Long ciliary nerve
Medulla
to orbicularis oculi muscles
Cornea

FIGURE V–19 Corneal blink reflex. When the cornea is touched, sensory signals project via V_1 to the spinal trigeminal nucleus. Second order sensory neurons project bilaterally to activate the lower motor neurons of the facial nuclei, which in turn activate the orbicularis oculi muscles to close the lids.

5. What is the blink reflex, and why is it important?

The blink reflex (see Fig. V–19) is the closure of the eyes in response to certain stimuli (bright light, corneal irritation, and loud noise). The motor component of the blink reflex (closing the eyes) is mediated by the branchial motor fibers of the CN VII (facial nerve). Several sensory pathways converge on the facial motor nucleus to activate the motor neurons. Initiating stimuli come from bright light via CN II, corneal stimulation via CN V (trigeminal), and loud sounds via CN VIII, auditory component.

Closing the eyes in response to intense light protects the retina from damage. Blinking in response to corneal irritation protects the cornea from airborne particles or other objects that could injure the eye. Blinking also prevents corneal dryness by moistening the cornea with tears. A dry cornea is painful and vulnerable to ulceration and infection. It is not certain why the eyes close in response to loud sounds, although presumably, loud sounds could predict flying debris.

CLINICAL TESTING

When testing the trigeminal nerve, it is important to remember to test both the sensory and the motor components. (See also “Cranial Nerves Examination” on the website.)

Sensory Component

The trigeminal nerve carries several sensory modalities, including discriminative touch, pain, temperature, and simple touch. All modalities are tested with the patient’s eyes closed. When testing, the examiner first must check bilaterally for the presence of each modality in the forehead (V_1), cheeks (V_2), and jaw (V_3) and determine whether both sides of the face are equally sensitive (Fig. V–20).

The discriminative touch pathway is tested by touching the skin (gently) with the sharp end of a pointed object (e.g., a toothpick) and asking the patient what he or she feels (see Fig. V–20). The pain and temperature pathway is tested by holding warm or cool objects against the skin. This can be done easily at the bedside by using the flat cool end of a tuning fork. Simple touch pathways are tested by lightly touching the skin with a wisp of cotton. The territories supplied by each division of the nerve are less variable in the central part of the face; therefore, testing should be done close to the midline.

The corneal reflex should always be tested when examining the trigeminal nerve. This test is particularly useful when assessing the integrity of V_1 in an unconscious patient. The corneal reflex is tested by observing whether the patient blinks in response to a light touch with a wisp of cotton on the cornea not the sclera (Fig. V–21).

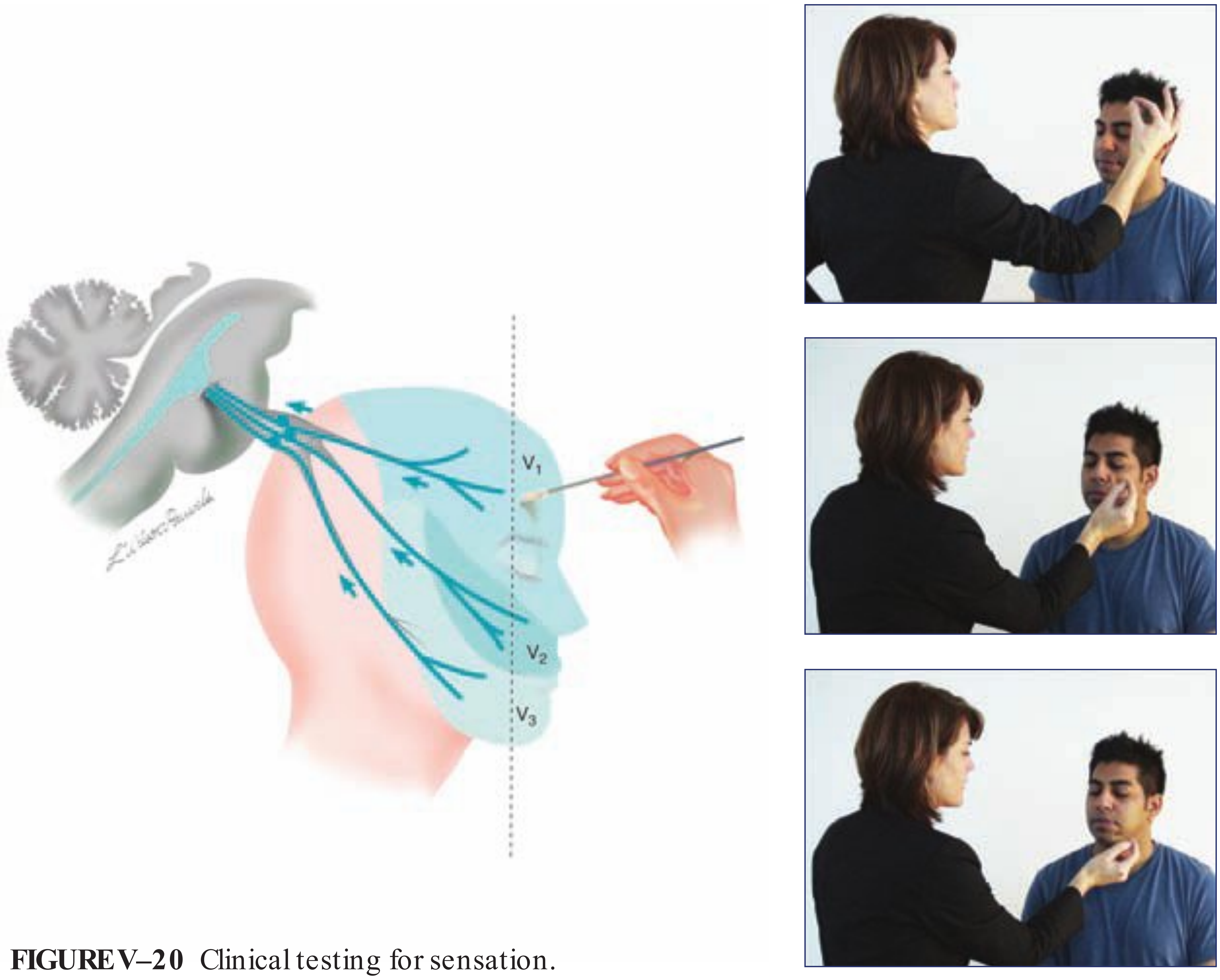

FIGURE V–20 Clinical testing for sensation.

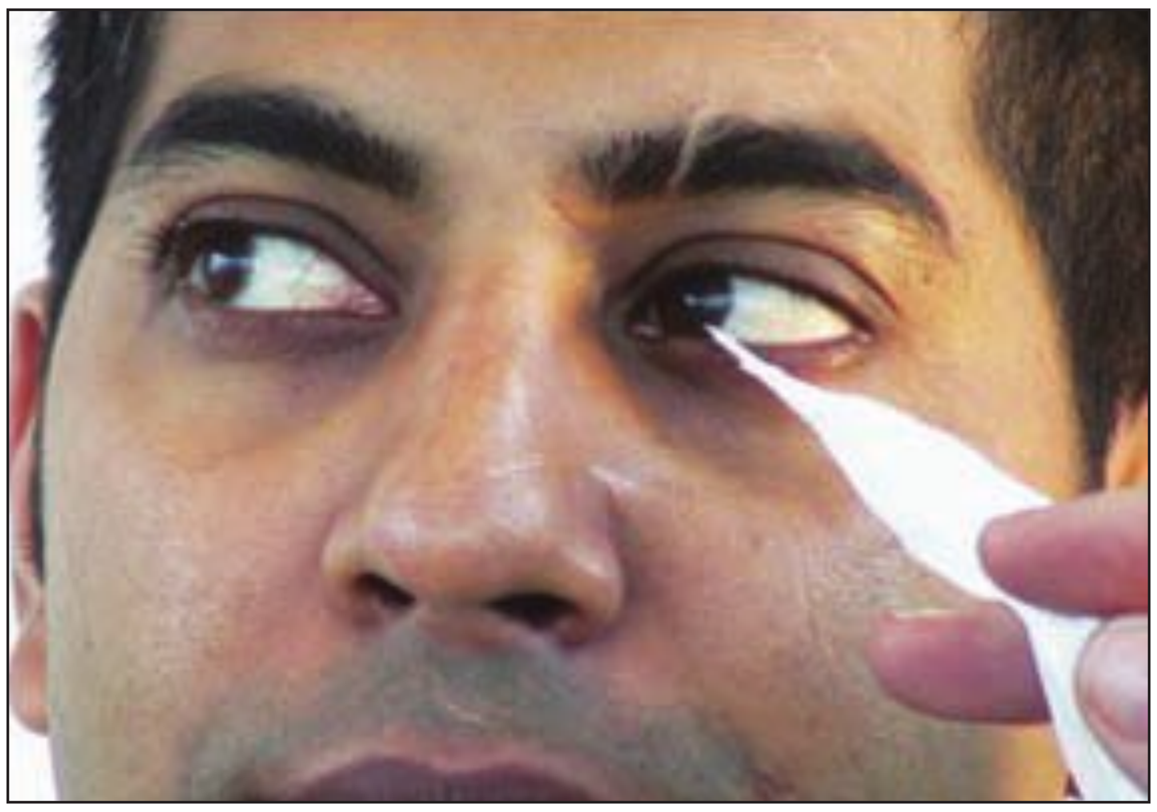

FIGURE V–21 Testing the corneal reflex.

Motor Component

To test the motor component of the trigeminal nerve, the examiner palpates the masseter and temporalis muscles on both sides and asks the patient to clamp the jaws tightly together. The examiner should feel the contraction of each muscle

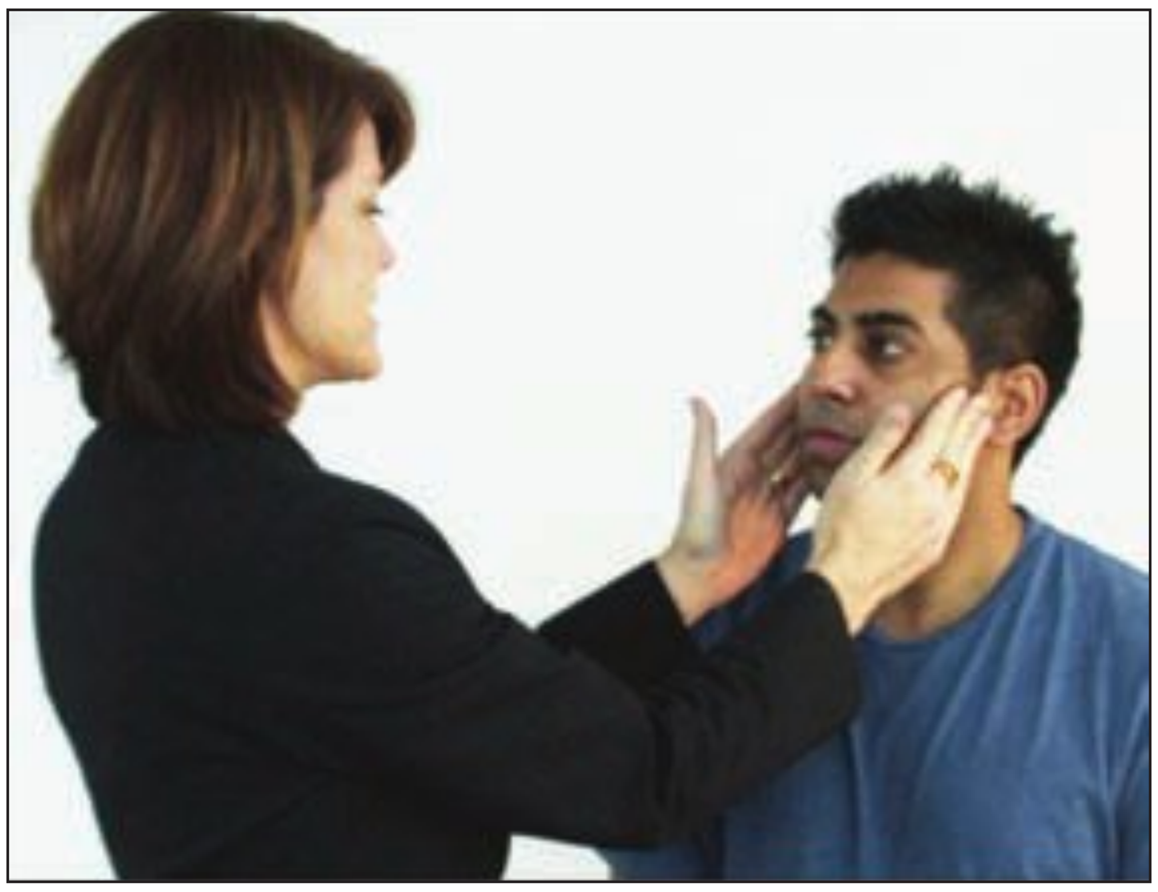

FIGURE V–22 Clinical testing for muscles of mastication.

(Fig. V–22). The patient is then asked to open her or his mouth so that the examiner can look for jaw deviation. If the motor component is not functioning correctly, the jaw will deviate toward the weak side. The patient is then asked to move the jaw to one side while the examiner attempts to push it back to the midline position. The examiner should not be able to overcome the strength of the pterygoid muscles. This is repeated on the other side.

To complete the motor examination, the jaw jerk should be tested. The mandibular (jaw jerk) reflex can be tested by tapping the middle of the chin with a reflex hammer while the patient's mouth is slightly open. A sudden slight closing of the jaw constitutes a normal reflex.

ADDITIONAL RESOURCES

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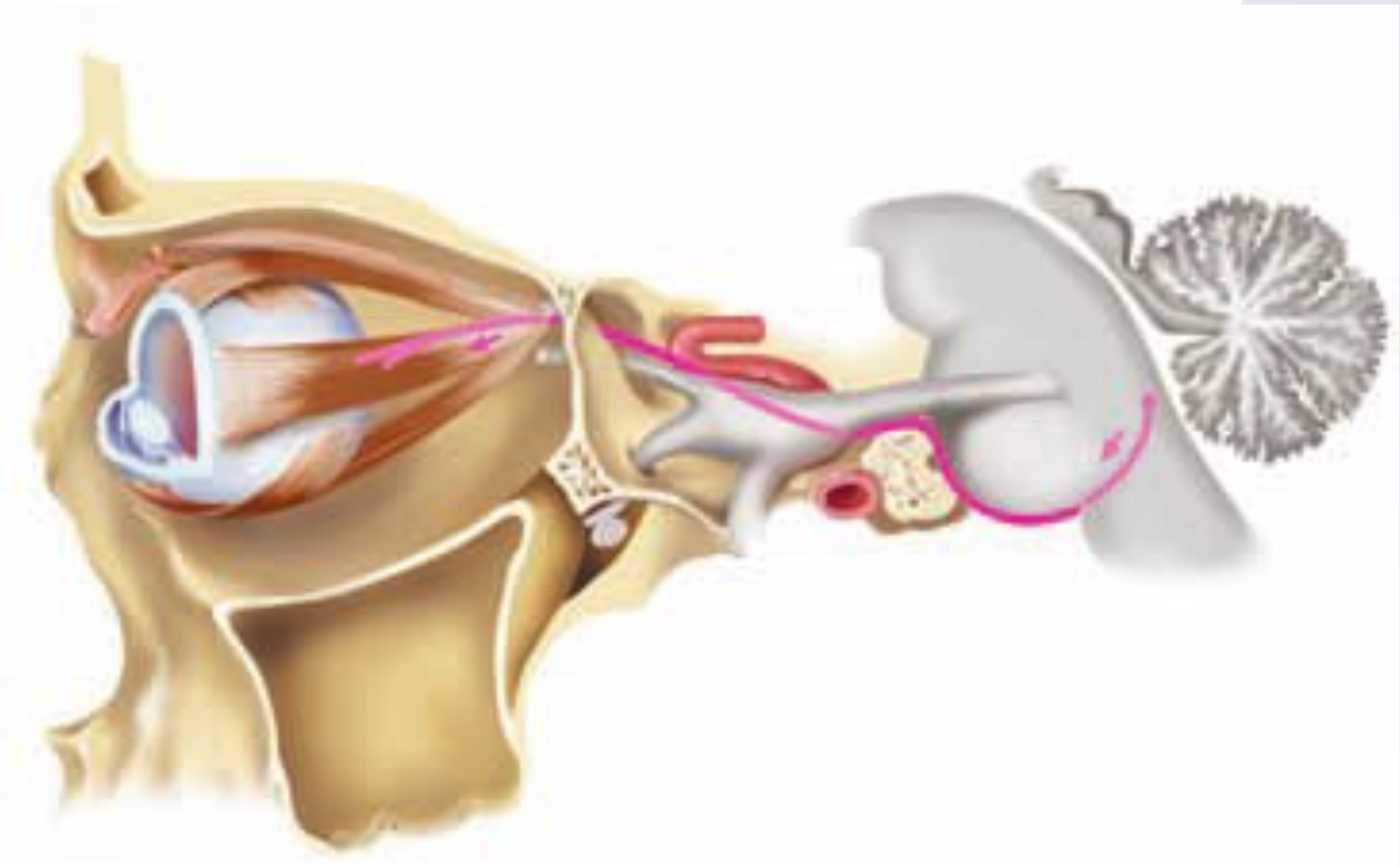

VI

Abducens Nerve

CASE HISTORY

Grace is a 36-year-old editor of a Hong Kong–based travel magazine. Grace had been well until about 3 months ago when she began having intermittent nosebleeds. The first nosebleed bled heavily for about 1 hour and stopped on its own. Since this first incident, Grace has had two further nosebleeds. About 2 months ago, she noticed that she had double vision when she looked to the left. Initially, it was intermittent, but it soon became a persistent problem. In the last month, she developed numbness of her left upper lip that spread to her left cheek and then to her forehead. Grace had been trying to ignore these symptoms, but when she had another bad nosebleed, she went to the emergency department of a nearby hospital.

Within 15 minutes of her arrival in the emergency department, Grace's nosebleed had stopped. She explained her recent symptoms to the doctor, who then called a neurologist. When the neurologist examined Grace, he found that her pupils were equal and reactive to light and that there was no evidence of ptosis (upper eyelid droop). The movements of her right eye were normal, but Grace clearly had problems moving her left eye. It was found that Grace was unable to abduct her left eye (look to the left), and when she attempted to do so, she would see a double image. However, she could look in all other directions without difficulty. When the neurologist tested for sensation on Grace's face, he found that she had numbness on her left forehead, cheek, and upper lip, while her lower lip and chin had normal sensation. Grace had symmetrical facial movements and there was no evidence of weakness of her facial muscles. The remainder of her cranial nerves (CNs) were functioning normally.

The doctor was concerned that Grace had a lesion involving her cavernous sinus, so he ordered a magnetic resonance imaging (MRI) scan of her brain. The scan demonstrated a mass from the pharynx infiltrating upward into the cavernous sinus. The doctor suspected that Grace might have a nasopharyngeal carcinoma and, therefore, arranged for an otolaryngologist to obtain a biopsy of the suspicious mass.

ANATOMY OF THE ABDUCENS NERVE

The abducens nerve has only a somatic motor component (Fig. VI–1). The function of this CN is to move the eye laterally away from the midline (Table VI–1) (the name is from the Latin words *ab*, away, and *ducere*, to lead). Axons of the abducens nerve emerge from the ventral aspect of the brain stem at the pontomedullary junction medial to CN VII. The nerve runs rostrally and slightly laterally in the subarachnoid space of the posterior cranial fossa to pierce the dura at a point lateral to the dorsum sellae of the sphenoid bone. It continues forward between the dura and the apex of the petrous temporal bone where it takes a sharp right-angled bend over the apex and enters the cavernous sinus (Fig. VI–2). Within the cavernous sinus, CN VI is situated lateral to the internal carotid artery and medial to CNs III, IV, V_1, and V_2 (Fig. VI–3). Continuing forward, the abducens nerve leaves the cavernous sinus and enters the orbit at the medial end of the superior orbital fissure. It passes through the tendinous ring and travels laterally to enter the deep surface of the lateral rectus muscle at about the midpoint of the muscle (Fig. VI–4).

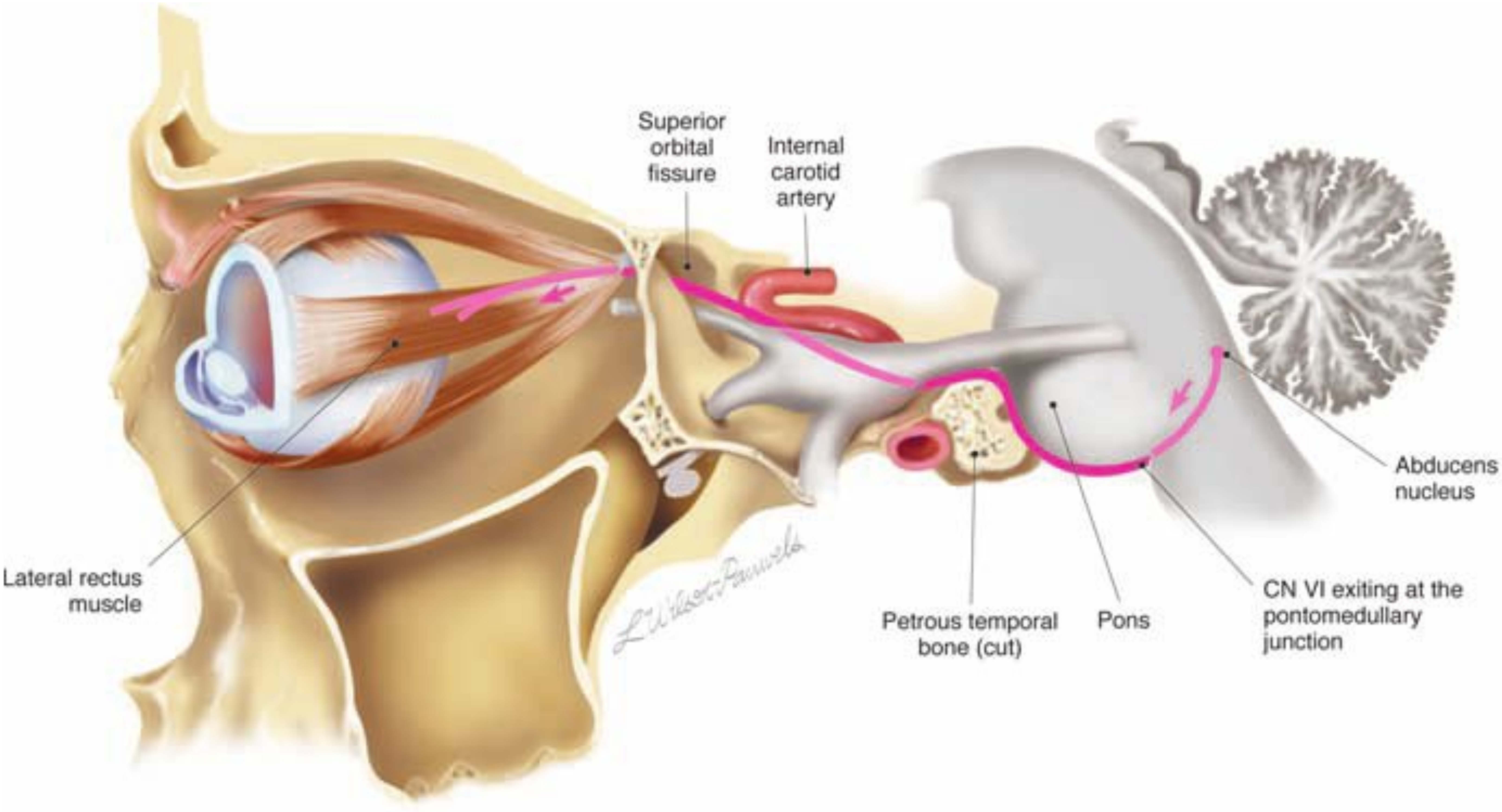

FIGURE VI–1 Overview of the abducens nerve.

TABLE VI–1 Component, Nucleus, and Function of the Abducens Nerve (CN VI)

Component	Nucleus	Function
Somatic motor (efferent)	Abducens nucleus	To innervate the lateral rectus muscle of the eye

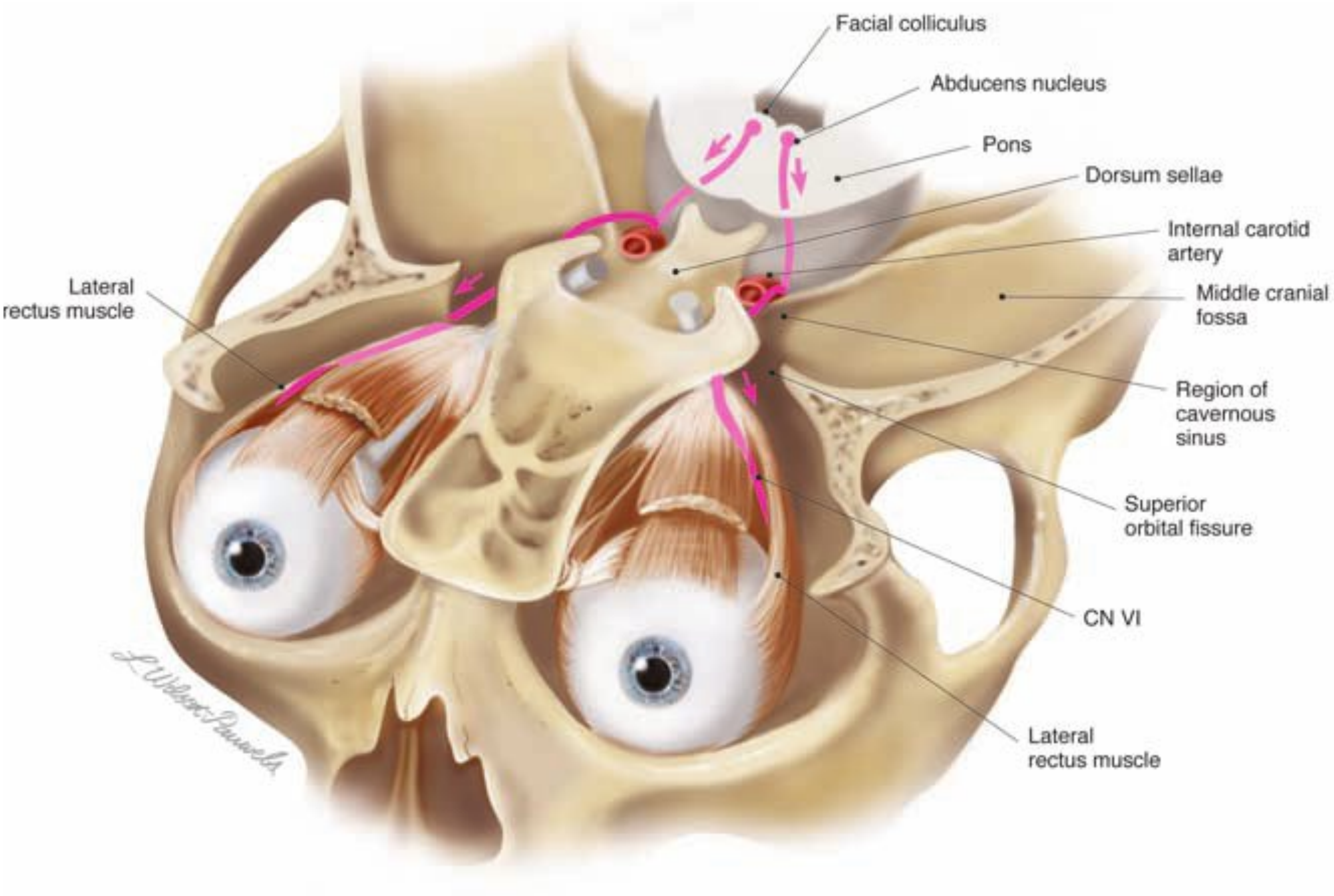

FIGURE VI–2 Route of the abducens nerve (cranial nerve VI) from the pons to the lateral rectus muscle.

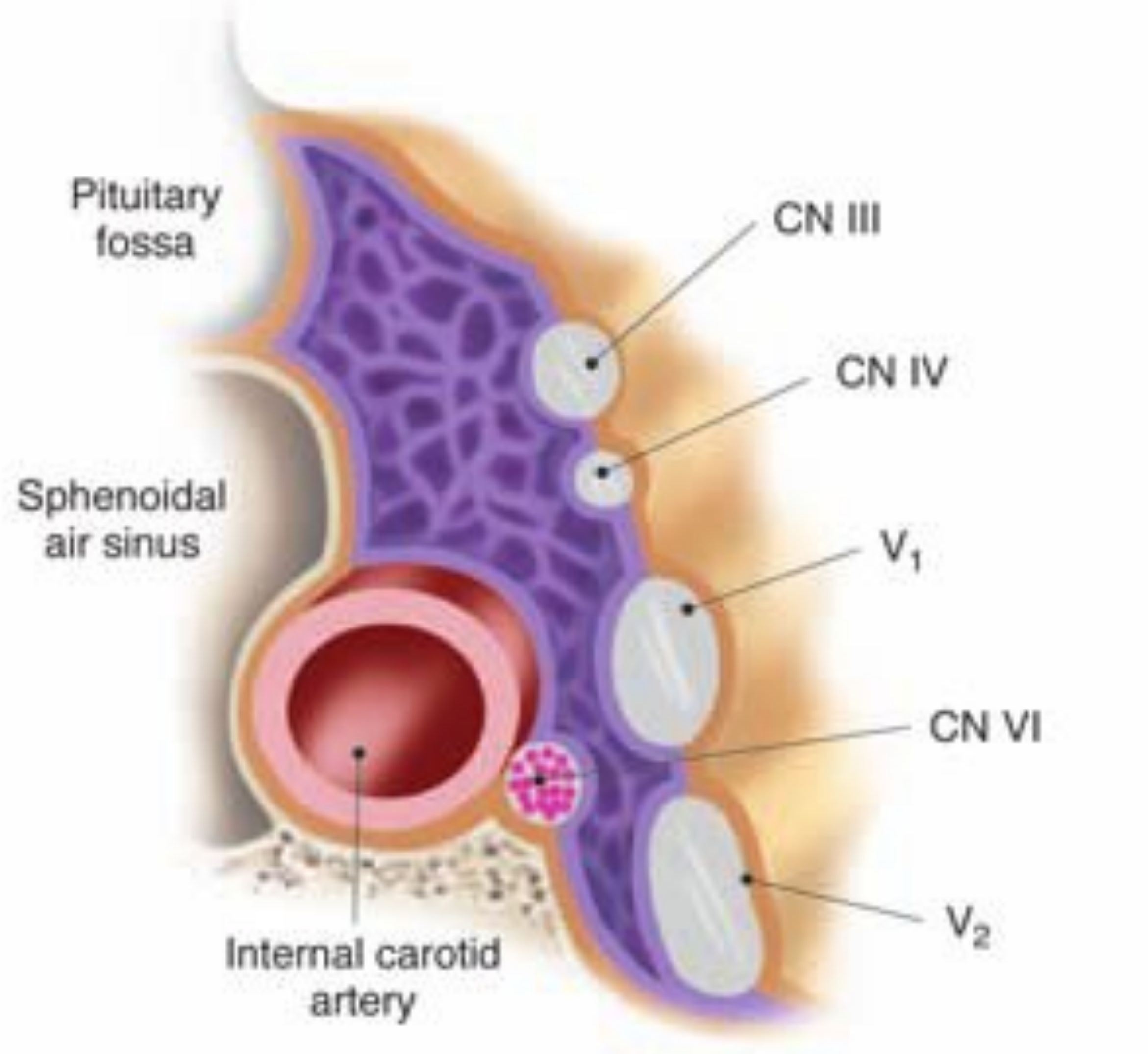

FIGURE VI–3 Slice through the left cavernous sinus showing the relationship of cranial nerve VI to other structures coursing through the sinus.

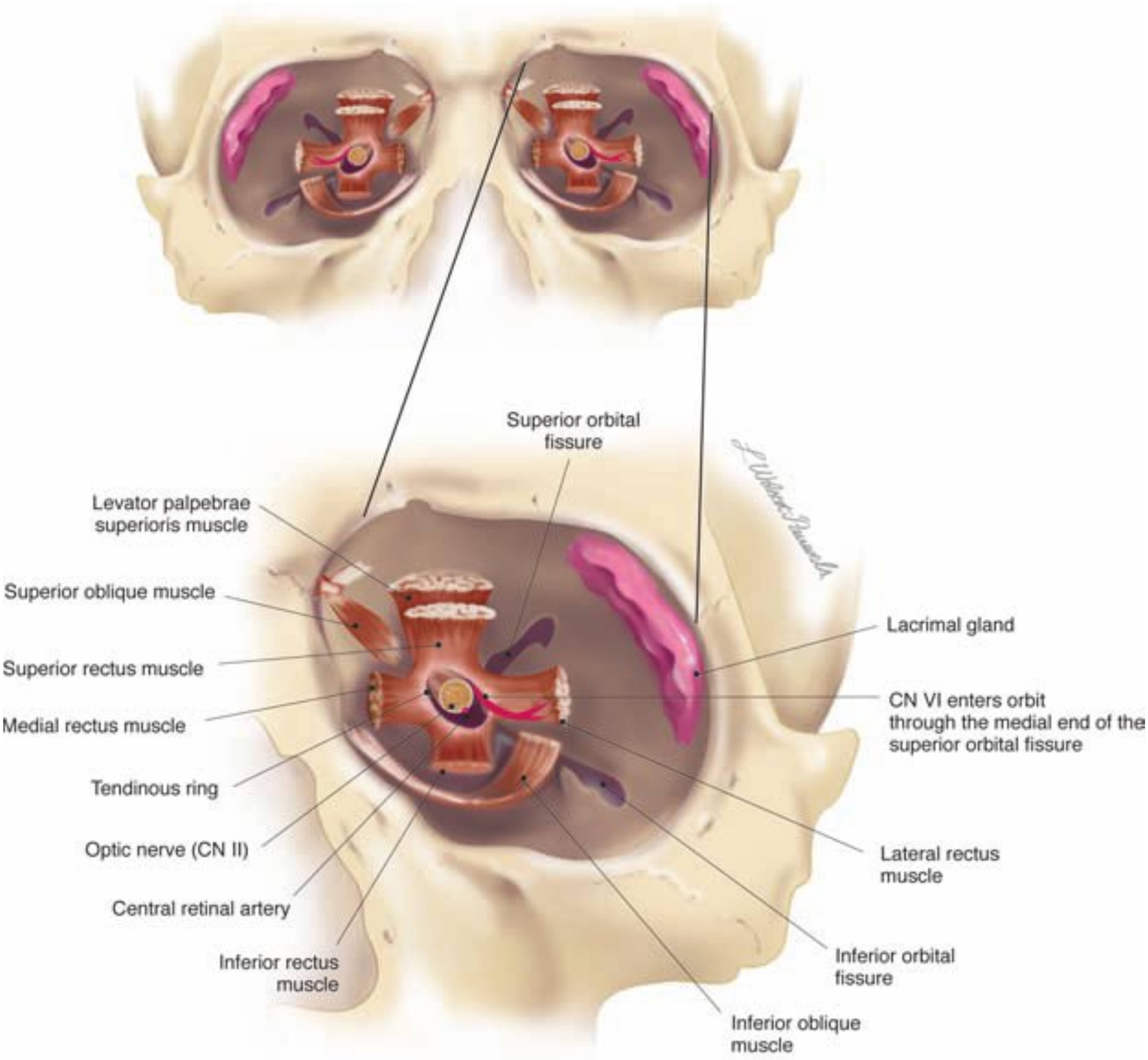

FIGURE VI–4 Apex of the right orbit illustrating the tendinous ring and the somatic motor component of cranial nerve VI.

The Abducens Nucleus

The abducens nucleus is located in the lower pons at the level of the facial colliculus. Like other somatic motor nuclei, the abducens nucleus is situated close to the midline. The nucleus is composed of two kinds of cells: lower motor neurons, whose axons constitute the abducens nerve, and internuclear neurons, whose axons ascend via the medial longitudinal fasciculus (MLF) to project to the medial rectus lower motor neurons in the contralateral oculomotor nucleus at the level of the superior colliculus (Fig. VI–5 inset). These are important for coordination of the lateral gaze (see Chapter 13). Axons of the lower motor neurons in the abducens nucleus course ventrally through the pons to emerge from the ventral surface of the brain stem at the pontomedullary junction (see Figs. VI–1 and VI–5).

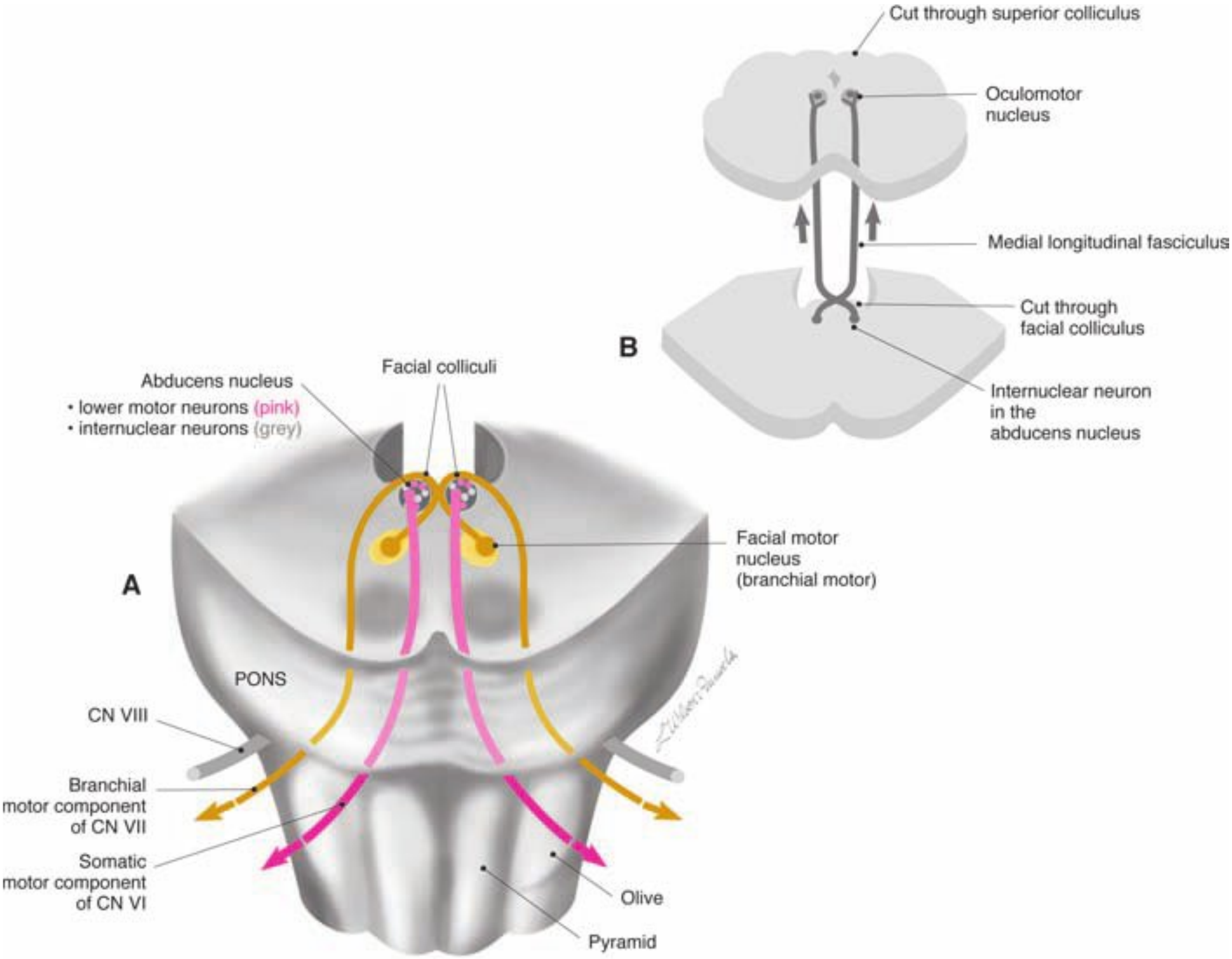
Cut through superior colliculus
Oculomotor nucleus
Medial longitudinal fasciculus
Cut through facial colliculus
B
Internuclear neuron in the abducens nucleus
Abducens nucleus
• lower motor neurons (pink)
• internuclear neurons (grey)
Facial colliculi
Facial motor nucleus (branchial motor)
A
PONS
CN VIII
Branchial motor component of CN VII
Somatic motor component of CN VI
Olive
Pyramid

FIGURE VI–5 Abducens nuclei in the brain stem.

A. Branchial motor axons from the facial nucleus loop over the abducens nucleus thereby creating an elevation (bump) in the floor of the fourth ventricle, called the facial colliculus. Because of this close anatomic association, lesions of the facial colliculus affect both cranial nerve VI and cranial nerve VII.

B. Insert illustrates the internuclear neurons ascending and crossing over to the contralateral oculomotor nucleus.

CASE HISTORY GUIDING QUESTIONS

1. Why was Grace having nosebleeds?
2. Why did Grace have double vision only when she was looking to the left?
3. How did the doctor know that the problem was within the cavernous sinus?
4. Why did Grace not have a numb chin?
5. Where along the course of CN VI could damage occur?

1. Why was Grace having nosebleeds?

The veins of the nose, including those of the highly vascular nasal mucosa, drain posteriorly into the cavernous sinus. The mass in Grace's posterior pharynx was invading her cavernous sinus, interfering with blood flow, and causing venous congestion in her nasal mucosa. The increased blood pressure within the delicate mucous membranes caused the small blood vessels to rupture and thus caused her nose to bleed.

2. Why did Grace have double vision only when she was looking to the left?

Horizontal eye movements require the coordinated action of the medial and lateral rectus muscles. The doctor noticed that when Grace attempted to look to the left, her left eye was unable to abduct (Fig. VI–6). This led him to conclude that Grace's left lateral rectus muscle was weak. On attempting leftward gaze, Grace was unable to align both eyes toward the same target. Therefore, the visual fields were projected onto different areas of the right and left retinae and two images were seen.

3. How did the doctor know that the problem was within the cavernous sinus?

The first step to solving this problem was to identify the nerves involved. Grace was unable to abduct her left eye. The lateral rectus muscle, which is responsible for eye abduction, is innervated by CN VI. Therefore, either her left lateral rectus muscle or her left CN VI was involved. Grace also had numbness over her left forehead and cheek. The ophthalmic division of the trigeminal nerve (V_1) supplies sensation to the forehead, the maxillary division of the trigeminal nerve (V_2) supplies sensation to the cheek, and the mandibular division of the trigeminal nerve (V_3) supplies

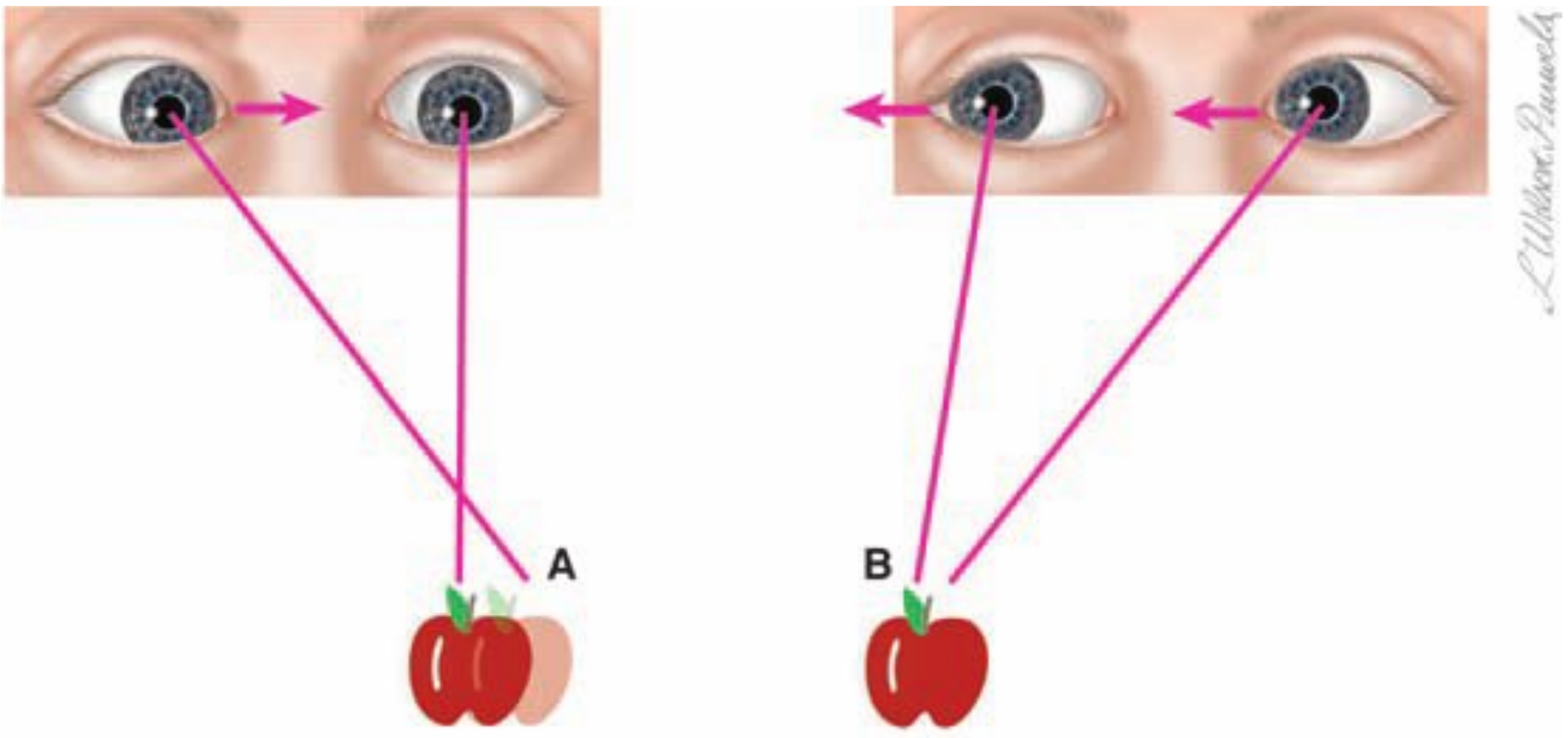

FIGURE VI–6 **A.** On attempted left lateral gaze, Grace was unable to abduct her left eye due to paralysis of her left lateral rectus muscle; therefore, she experienced double vision. **B.** When looking to the right, Grace was able to direct both eyes toward the same object.

sensation to the chin (see Chapter V). Therefore, the left V_1 and V_2 were also affected but the left V_3 was spared.

The second step to solving the problem involves a knowledge of the course of the CNs. It is reasonable to assume there is only one cause for Grace's problem and, therefore, that the site of the lesion is located where CN VI and CNs V_1 and V_2 are in close proximity to each other. The only anatomic place where this occurs is within the cavernous sinus (see Fig. VI–3). Therefore, disease processes of the cavernous sinus can affect these nerves.

4. Why did Grace not have a numb chin?

The mandibular division of the trigeminal nerve (V_3) passes just outside the cavernous sinus and, therefore, was not involved by the infiltrating mass.

5. Where along the course of CN VI could damage occur?

CN VI can be damaged anywhere along its course from its nucleus in the pons to its target muscle in the orbit (the lateral rectus muscle).

- Within the Brain Stem

  The nucleus and its axons can be affected by infarction, tumor, or demyelination.
- Between the brain stem and the cavernous sinus

  The nerve itself can become ischemic (this is most commonly seen in patients with diabetes or hypertension). The nerve can be affected by meningeal infection. Occasionally, cerebellopontine angle tumors can involve CN VI as well as CNs V, VII, and VIII (see Chapter VIII, Fig. VIII–3 check later). Mastoiditis (inflammation of the mastoid process of the temporal bone) can involve CN V as well as CN VI (Gradenigo's syndrome).
- Within the cavernous sinus

  As CN VI passes anteriorly through the cavernous sinus, the nerve can be involved in any disease process within the sinus (carotid aneurysm, sinus thrombosis, infection, inflammation, or tumor). Other CNs are commonly affected at this site (III, IV, V_1, V_2) owing to their close proximity within the cavernous sinus. Grace was found to have a nasopharyngeal carcinoma invading the cavernous sinus involving the left CN VI, V_1 and V_2.
- Within the superior orbital fissure

  CN VI passes anteriorly through the superior orbital fissure to reach the lateral rectus muscle. Fractures of the orbit or orbital tumors could compromise the nerve. Other CNs (III, IV, and V_1) passing through the fissure also could also be affected.
- Within the orbit

  CN VI could be injured by an orbital fracture as it passes through the orbit to innervate the lateral rectus muscle.

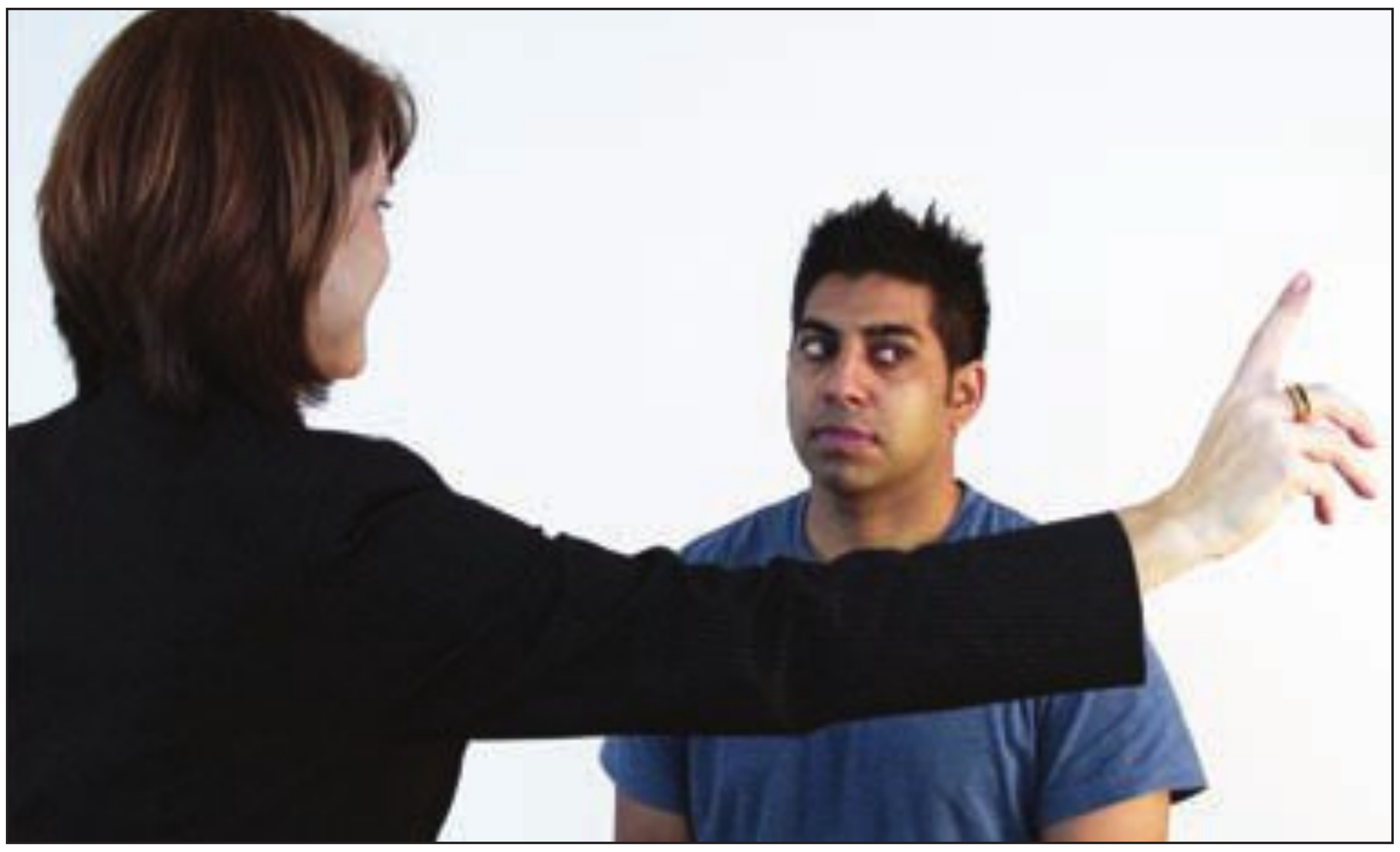

FIGURE VI–7 Testing the abducens nerve.

CLINICAL TESTING

CN VI is tested in conjunction with CNs III and IV through the assessment of eye movements (see Chapter 13). When testing specifically for CN VI, it is important to bring the eye through the full extent of the horizontal plane and to ensure that the eye moves fully away from the midline (Fig. VI–7). The iris should bury itself into the palpebral fissure of the corner of the eye and the white of the sclera should not be seen. (See also "Cranial Nerves Examination" on website.)

ADDITIONAL RESOURCES

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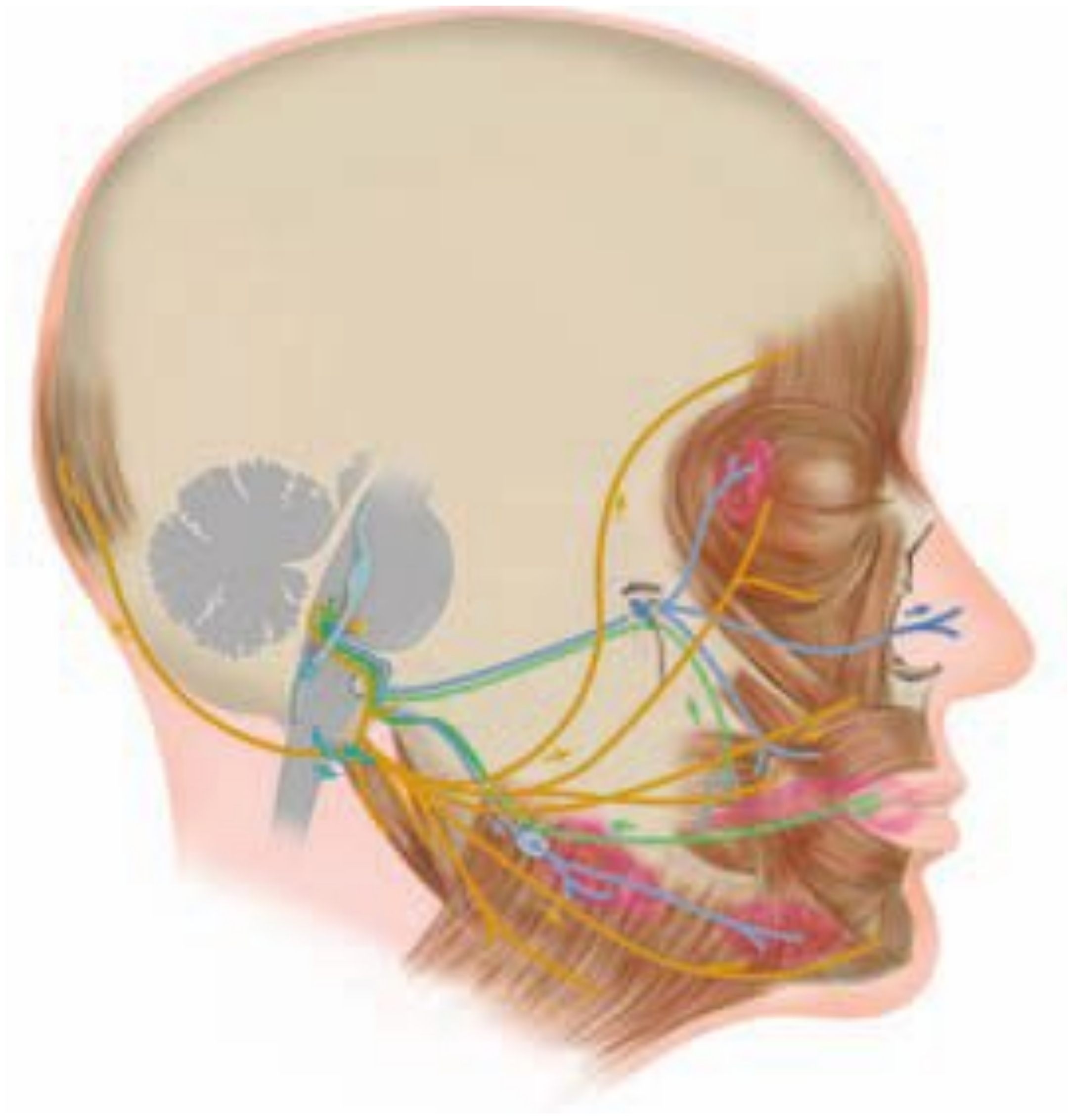


VII

Facial Nerve

CASE HISTORY

John is a 45-year-old man in the prime of his career. One morning he was having some difficulties shaving that resulted in a couple of nicks on the right side of his face. He went to work and, at lunch time, ordered soup because his mouth felt unusually dry. To his embarrassment, the soup dribbled out of the right corner of his mouth. After lunch, he looked in the mirror and noticed that the entire right side of his face was drooping, but otherwise, his face felt normal. John phoned his family doctor who arranged to see him that afternoon.

At the end of the day when the family doctor saw John, he was unable to raise his right eyebrow or completely close his right eye. Nor was he able to raise the right corner of his mouth, and as a result, his face appeared to be pulled to the left. The doctor also discovered that John had lost the ability to taste anything on the anterior two-thirds of his tongue on the right side. There was no change in sensation on his face, but John perceived sounds to be louder in his right ear. John also had a lot of tearing of his right eye. The rest of John's cranial nerves were functioning normally. John's doctor diagnosed Bell's palsy, assured him that he should make a good recovery, and started him on an appropriate medication.

Despite expectations, 6 weeks later, there was no apparent change, but by 8 months after the onset, the right side of John's face was almost back to normal. John saw his doctor again, who observed an interesting phenomenon. Every time John smiled, his right eye would close. There were no other apparent abnormalities and John was reassured by his doctor that this unusual motor activity was a consequence of the Bell's palsy and that he was otherwise normal.

ANATOMY OF THE FACIAL NERVE

The facial nerve is the most frequently compromised of all the cranial nerves (CNs) owing to its long course through the head. Figure VII–1 provides a diagrammatic overview of the nerve and Table VII–1 lists its components, nuclei, ganglia, and functions.

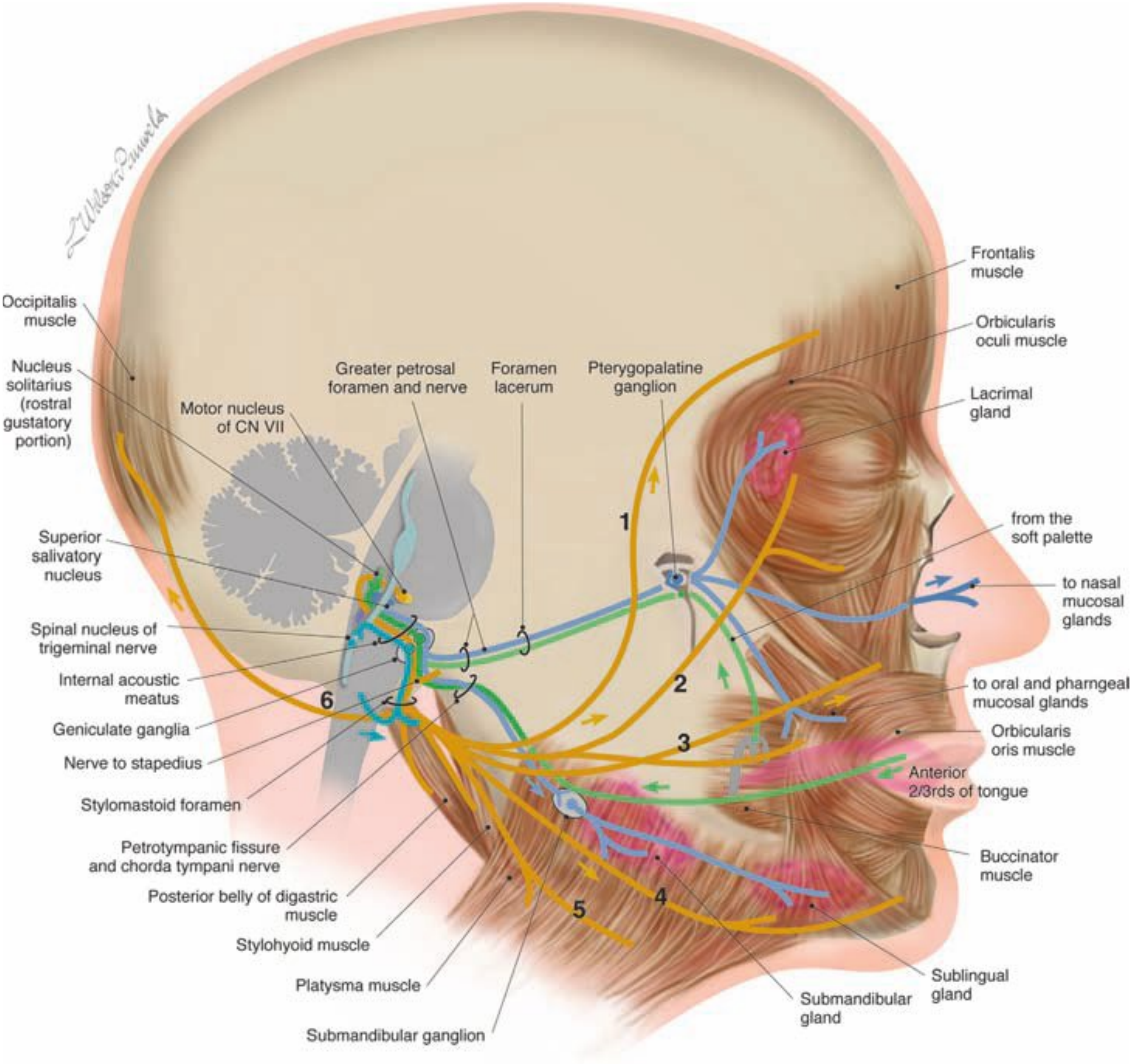


FIGURE VII–1 Overview of facial nerve components (parotid gland removed).

MAJOR BRANCHES:
1. Temporal nerve
2. Zygomatic nerve
3. Buccal nerve
4. Mandibular nerve
5. Cervical nerve
6. Posterior auricular nerve

TABLE VII–1 Components, Nuclei, Ganglia and Functions of the Facial Nerve (CN VII)

Component	Nucleus	Ganglion and Cell of Origin	Function
General sensory (afferent)	Pontine trigeminal nucleus (for touch) Spinal trigeminal nucleus (for pain)	Geniculate ganglion	For general sensation from a small variable area of the concha of the external ear (pinna/auricle), external acoustic meatus, and external (lateral) surface of the tympanic membrane, and a small area of skin behind the ear
Special sensory (afferent)	Nucleus solitarius (rostral gustatory portion)	Geniculate ganglion Taste buds	For taste sensation from the anterior two-thirds of the tongue and soft palate
Branchial motor (efferent)	Motor nucleus of cranial nerve VII		To innervate the muscles of facial expression (see Table VII–2 for more detail)
Parasympathetic motor (visceral efferent)	Superior salivatory nucleus	Pterygopalatine and submandibular ganglia	To stimulate the lacrimal, submandibular, and sublingual glands as well as the oral, nasal, and pharyngeal mucosal glands

CENTRAL COMPONENTS OF THE FACIAL NERVE

The branchial motor component forms the largest part of CN VII. The neuronal cell bodies that form he facial nucleus are situated in the pons. Their axons course dorsally over the nucleus of CN VI, forming the internal genu, and then ventrally to emerge from the brain stem at the pontomedullary junction. The remaining components, the general sensory, special sensory, and parasympathetic motor, relate to the trigeminal nucleus, the gustatory nucleus (rostral part of nucleus solitarius), and the superior salivatory nucleus, respectively. Their axons emerge from the brain stem at the pontomedullary junction lateral to the branchial motor axons. They are bound in a common sheath separate from the branchial motor components and are called the “nervus intermedius” (Fig. VII–2). Within the internal acoustic meatus, the nervus intermedius joins with the branchial motor axons.

Course of the Facial Nerve

Both components of CN VII emerge from the brain stem at the pontomedullary junction, cross the subarachnoid space, and enter the internal acoustic meatus (Fig. VII–3; see also Fig. VII–1). At the lateral end of the meatus, the facial nerve enters its own canal, named the “facial canal,” and runs laterally, above the vestibule (part of the osseous labyrinth of the inner ear), and bends sharply backward. This sharp

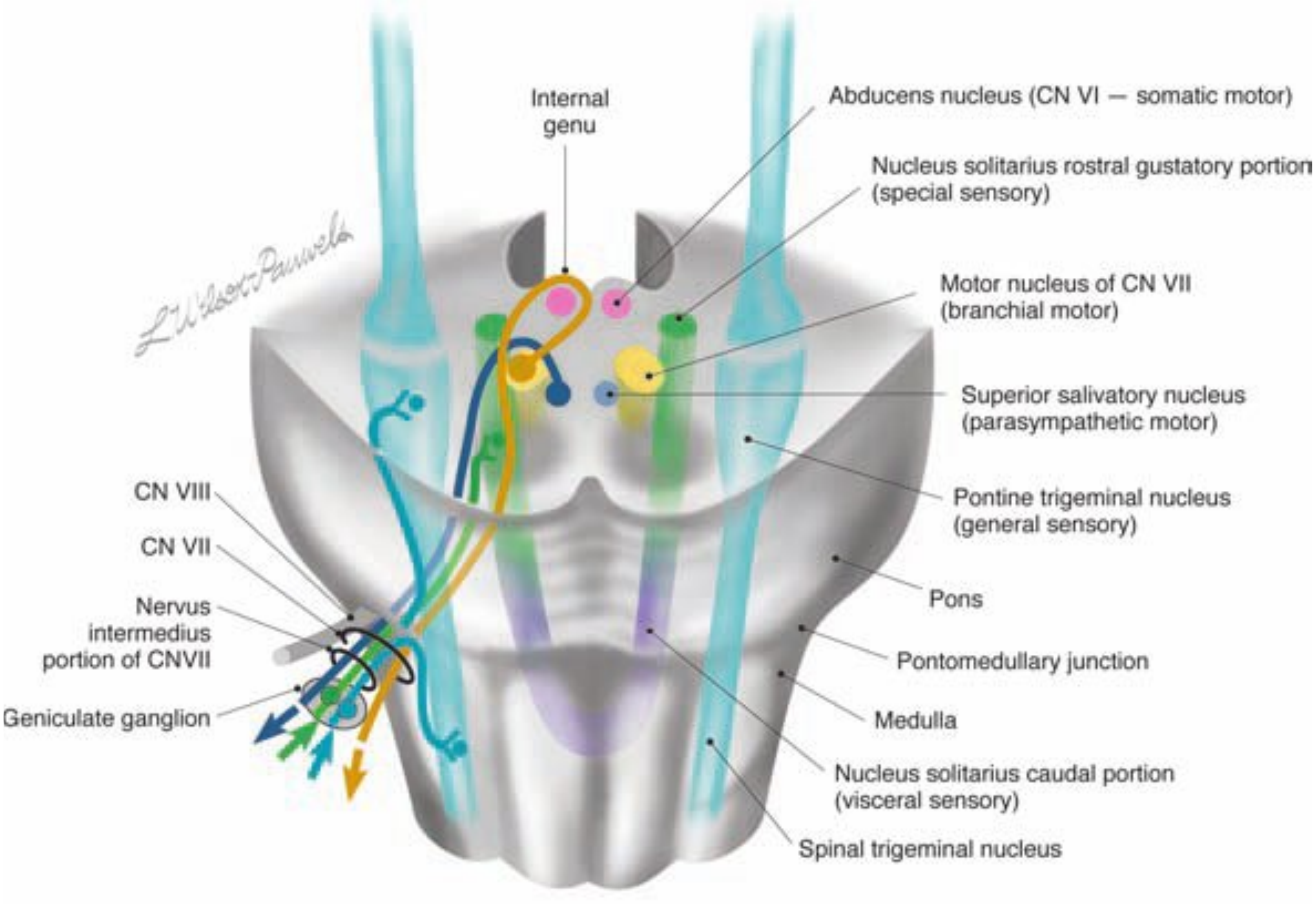

Internal genu
Abducens nucleus (CN VI — somatic motor)
Nucleus solitarius rostral gustatory portion (special sensory)
Motor nucleus of CN VII (branchial motor)
Superior salivatory nucleus (parasympathetic motor)
Pontine trigeminal nucleus (general sensory)
Pons
Pontomedullary junction
Medulla
Nucleus solitarius caudal portion (visceral sensory)
Spinal trigeminal nucleus
CN VIII
CN VII
Nervus intermedius portion of CNVII
Geniculate ganglion

FIGURE VII–2 Cranial nerve VII leaving brain stem nuclei at pontomedullary junction (fibers spread apart).

bend is called the “geniculum” and is the site of the geniculate ganglion where the cell bodies of the general and special sensory neurons are located. At the geniculate ganglion, the facial nerve gives off the greater petrosal nerve, which carries parasympathetic signals to the lacrimal gland, oral, pharyngeal, and nasal mucosal glands and taste signals from the soft palate. The remaining axons then continue along the facial canal, across the medial wall of the aditus to the mastoid antrum, and down the posterior wall of the tympanic cavity to the stylomastoid foramen.

Two nerves branch from the main trunk as it passes through the vertical part of the facial canal. Uppermost, the nerve to the stapedius muscle enters directly into the base of the muscle. Below it, the chorda tympani branches off and courses through the middle ear cavity, carrying taste sensations in from the anterior two-thirds of the tongue and parasympathetic motor signals out to the sublingual and submandibular salivary glands.

The remaining branchial motor and general sensory axons emerge from the stylomastoid foramen. Motor branches are given off to the stylohyoid, posterior belly of digastric and occipitalis muscles. The remaining branchial motor axons pass forward through the substance of the parotid gland to supply the muscles of facial expression (see Fig. VII–1). A small group of sensory axons from the posterior auricular branch carrying general sensations from the concha of the external ear, external acoustic meatus, and tympanic membrane (Fig. VII–4) enter the stylomastoid foramen.

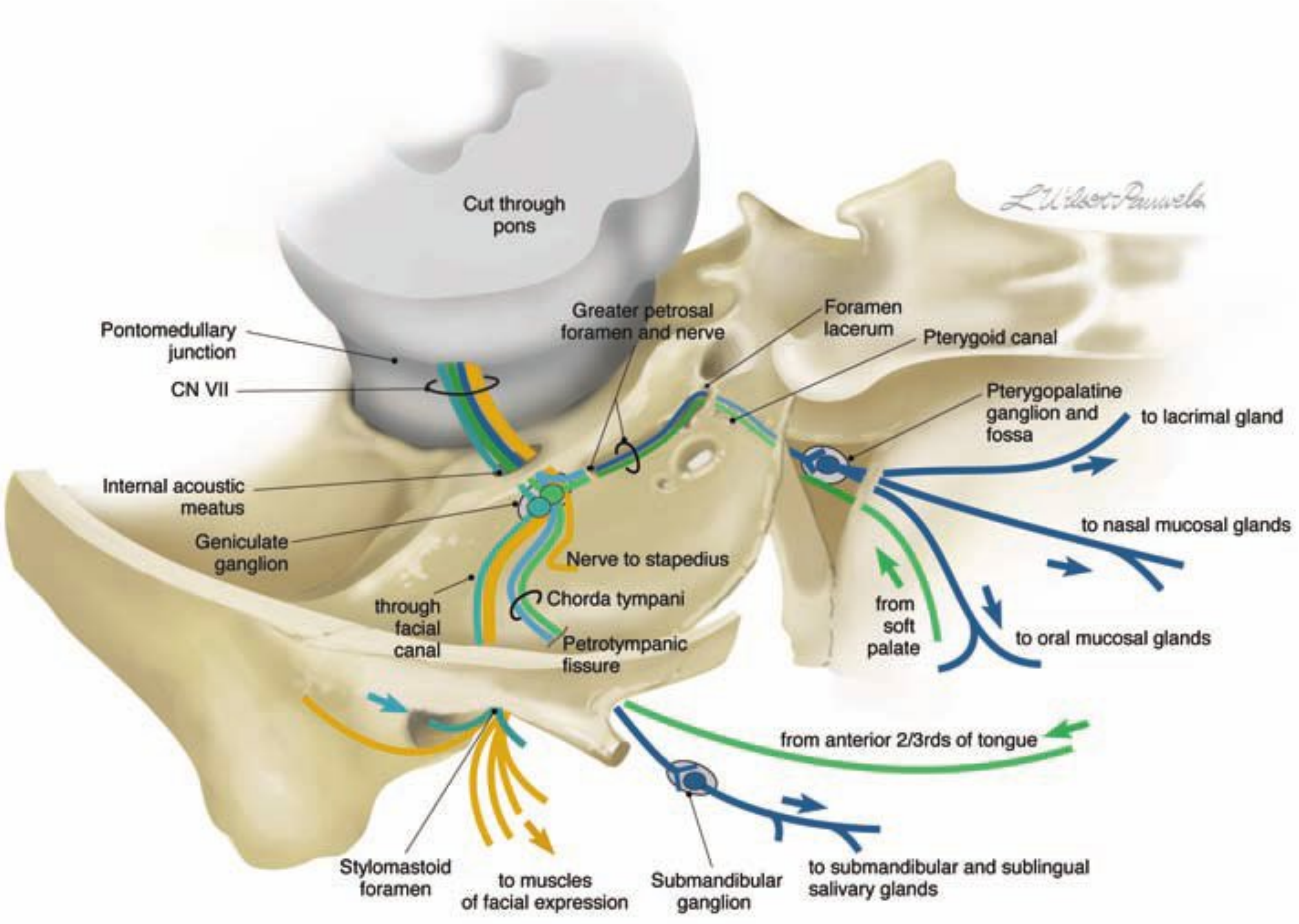


FIGURE VII–3 Superior view of the route of cranial nerve VII from the pons through the cranium (brainstem is elevated).

GENERAL SENSORY (AFFERENT) COMPONENT

The general sensory component of CN VII is very small (see Fig. VII–4). It provides a variable contribution to CNs V and X in innervating the concha of the external ear, the external acoustic meatus, and the external (lateral) surface of the tympanic membrane. The nerve cell bodies are located in the geniculate ganglion, and their central axons form part of nervus intermedius.

Touch signals enter the brain stem and project to the pontine trigeminal nucleus and, via the medial lemniscus, to the ventral posterior nucleus of the thalamus and on to the sensory cortex. Pain signals descend and project to the spinal trigeminal nucleus and, via the spinal lemniscus, to two distinct populations of neurons in the thalamus: neurons which project to the sensory cortex for identification of the location and intensity of the pain, and neurons which project to the cingulate cortex where the emotional component of pain is mediated.

It is thought that only pain signals that arise from the tympanic membrane are carried in CN VII.

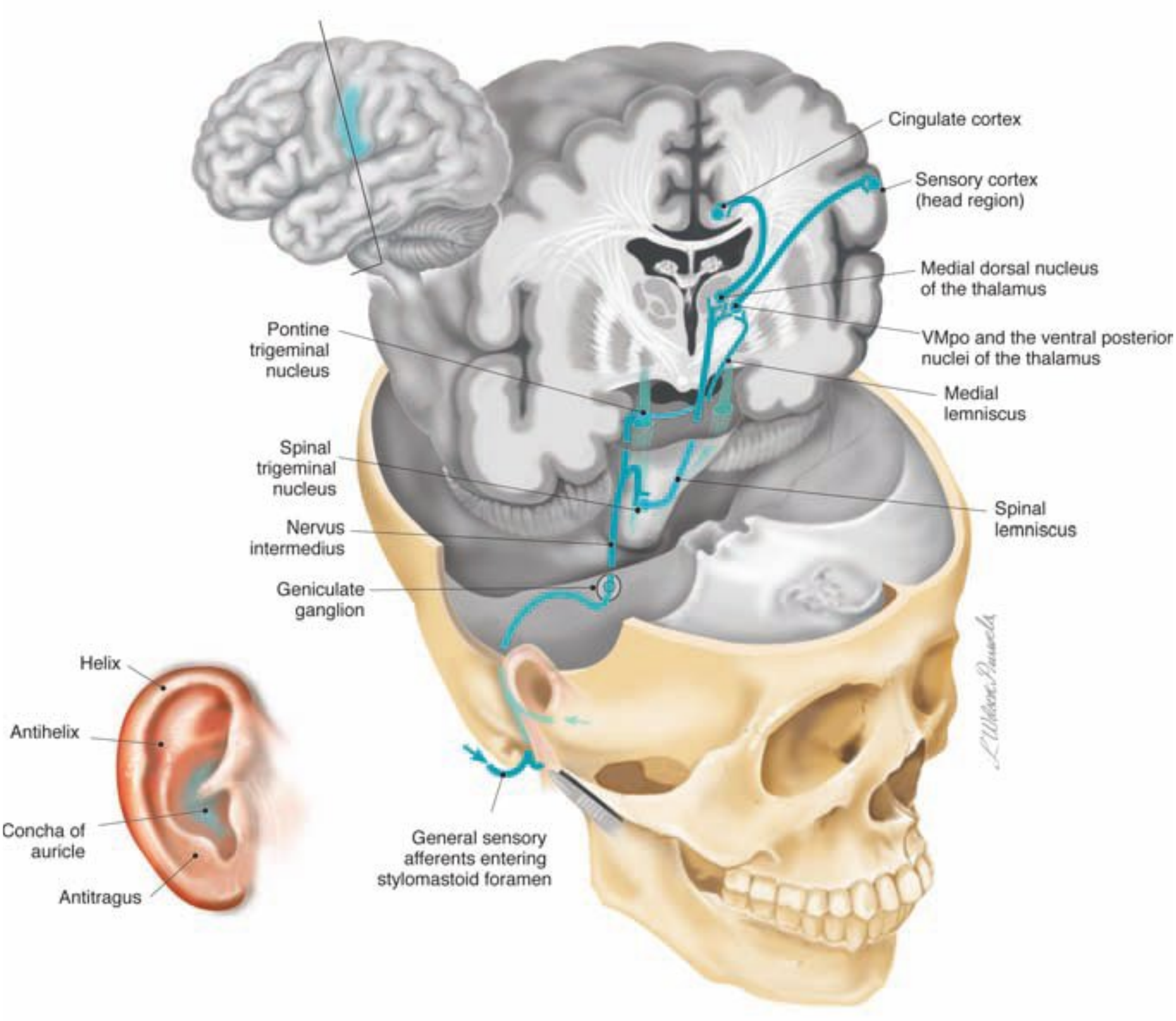
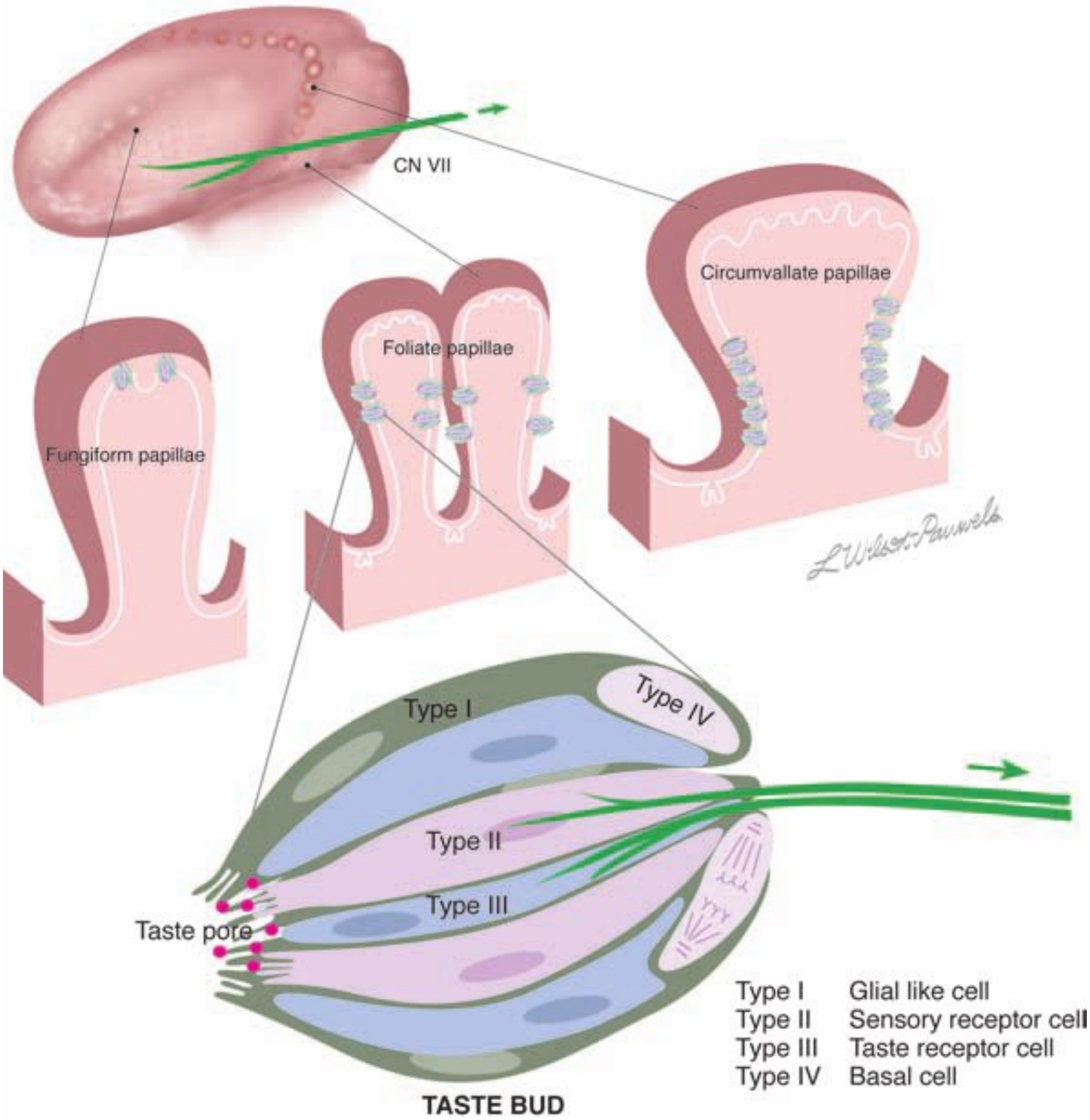

Cingulate cortex
Sensory cortex (head region)
Medial dorsal nucleus of the thalamus
VMpo and the ventral posterior nuclei of the thalamus
Medial lemniscus
Spinal lemniscus
Pontine trigeminal nucleus
Spinal trigeminal nucleus
Nervus intermedius
Geniculate ganglion
Helix
Antihelix
Concha of auricle
Antitragus
General sensory afferents entering stylomastoid foramen
L.Wilson-Pauwels

FIGURE VII–4 General sensory (pain and touch) component of the facial nerve.

SPECIAL SENSORY (AFFERENT) COMPONENT

Human beings have approximately 5000 taste buds located predominantly on the tongue and soft palate. On the tongue, they are associated with fungiform, circumvallate, and foliate papillae (Fig. VII–5). They form clusters of 1200 to 1600 cells arranged like garlic cloves within a garlic bud. Taste cells, like olfactory receptor cells, are replicated throughout life, with a generation time of approximately 10 days. There are five basic tastes: salt, sour, sweet, bitter, and savory (umami).

The special sensory axons of CN VII carry signals from taste buds on the anterior two-thirds of the tongue via the chorda tympani nerve and from a small population of taste buds on the soft palate via the greater petrosal nerve (Fig. VII–6). Their cell bodies are located in the geniculate ganglion within the facial canal.



Type I	Glial like cell
Type II	Sensory receptor cell
Type III	Taste receptor cell
Type IV	Basal cell

FIGURE VII–5 Taste buds

Axons from the tongue form part of the lingual nerve, and then separate from it to join the chorda tympani nerve, which passes through the petrotympanic fissure and the middle ear cavity medial to the tympanic membrane to join the facial nerve in the facial canal.

Axons from the soft palate course through the pterygopalatine fossa and foramen lacerum to become part of the greater petrosal nerve, which enters the greater petrosal foramen to reach the geniculate ganglion.

From the ganglion, all special sensory axons enter the brain stem at the pontomedullary junction as part of the nervus intermedius. They then enter the tractus solitarius in the brain stem and synapse in the rostral (gustatory) part of the nucleus solitarius. Unlike other sensory projections, the ascending (secondary) fibers from

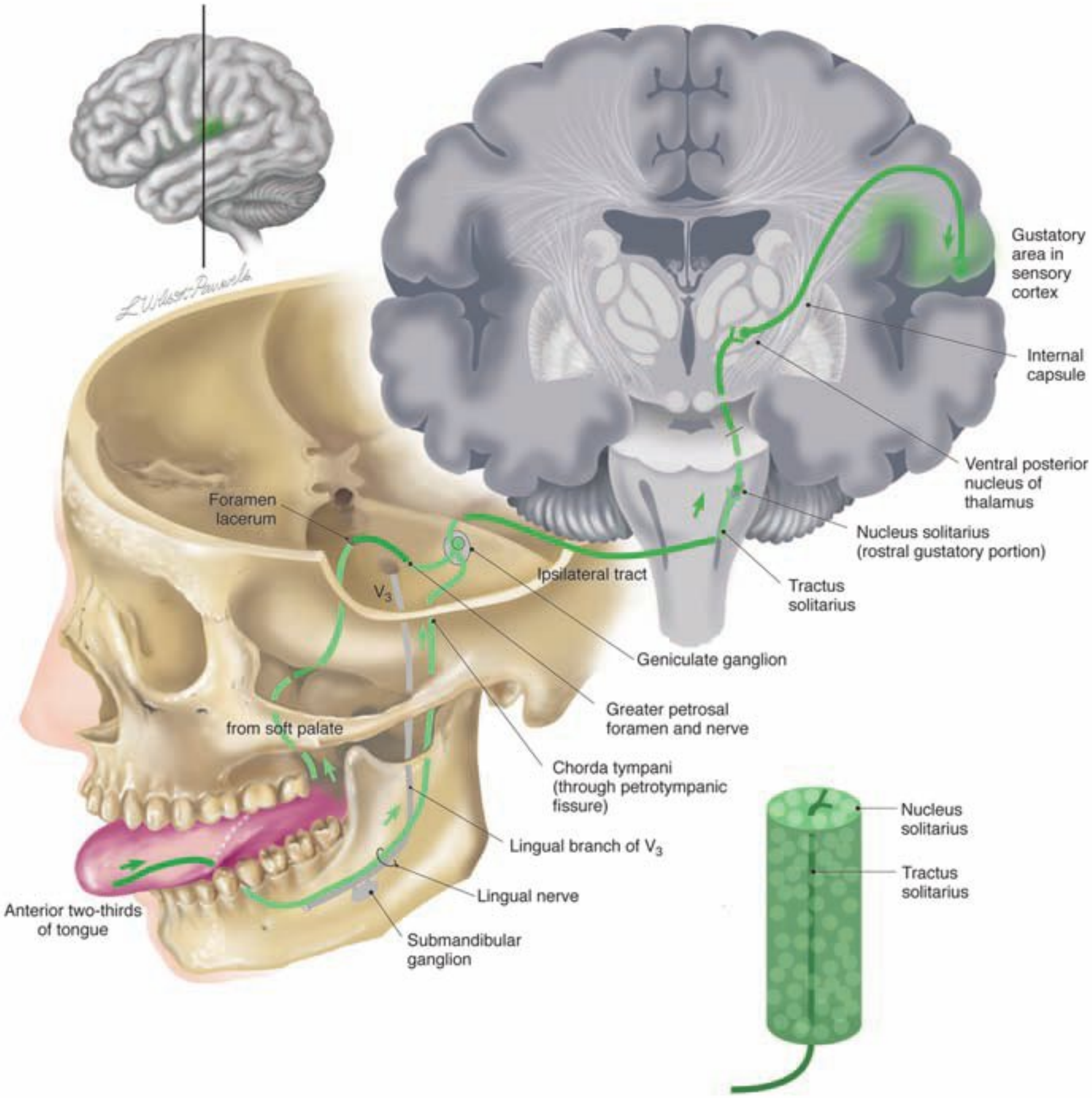

Gustatory area in sensory cortex
Internal capsule
Ventral posterior nucleus of thalamus
Nucleus solitarius (rostral gustatory portion)
Foramen lacerum
Ipsilateral tract
Tractus solitarius
V_3
Geniculate ganglion
Greater petrosal foramen and nerve
from soft palate
Chorda tympani (through petrotympanic fissure)
Lingual branch of V_3
Lingual nerve
Anterior two-thirds of tongue
Submandibular ganglion
Nucleus solitarius
Tractus solitarius

FIGURE VII–6 Special sensory component of the facial nerve

this nucleus project *ipsilaterally* to reach the ventral posterior nucleus of the thalamus. Axons of thalamic (tertiary) neurons then project through the posterior limb of the internal capsule to the cortical area for taste, which is located in the most inferior part of the sensory cortex in the postcentral gyrus and extends onto the insula (see Fig. VII–6).

BRANCHIAL MOTOR (EFFERENT) COMPONENT

Signals for voluntary movements of the facial muscles originate in the motor areas of the cerebral cortex. They travel through the posterior limb of the internal capsule as part of the corticobulbar tract and project to the ipsilateral and contralateral motor nuclei of CN VII in the tegmentum of the caudal pons (Fig. VII–7).

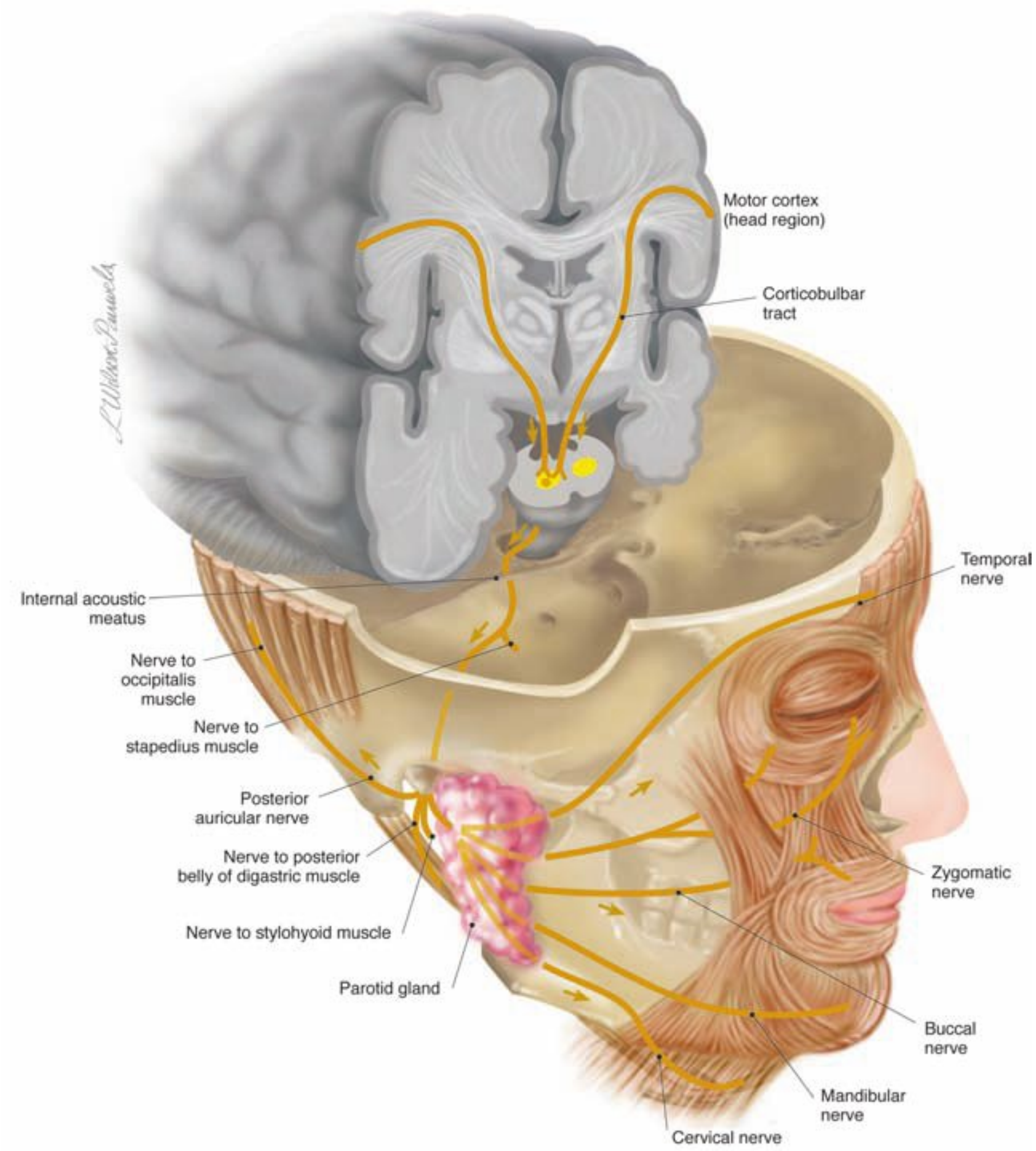

Motor cortex (head region)
Corticobulbar tract
Temporal nerve
Internal acoustic meatus
Nerve to occipitalis muscle
Nerve to stapedius muscle
Posterior auricular nerve
Nerve to posterior belly of digastric muscle
Nerve to stylohyoid muscle
Parotid gland
Zygomatic nerve
Buccal nerve
Mandibular nerve
Cervical nerve

FIGURE VII–7 Branchial motor component of the facial nerve demonstrating ipsilateral and contralateral innervation of the right facial muscles (cortex and brainstem are elevated and turned anteriorly).

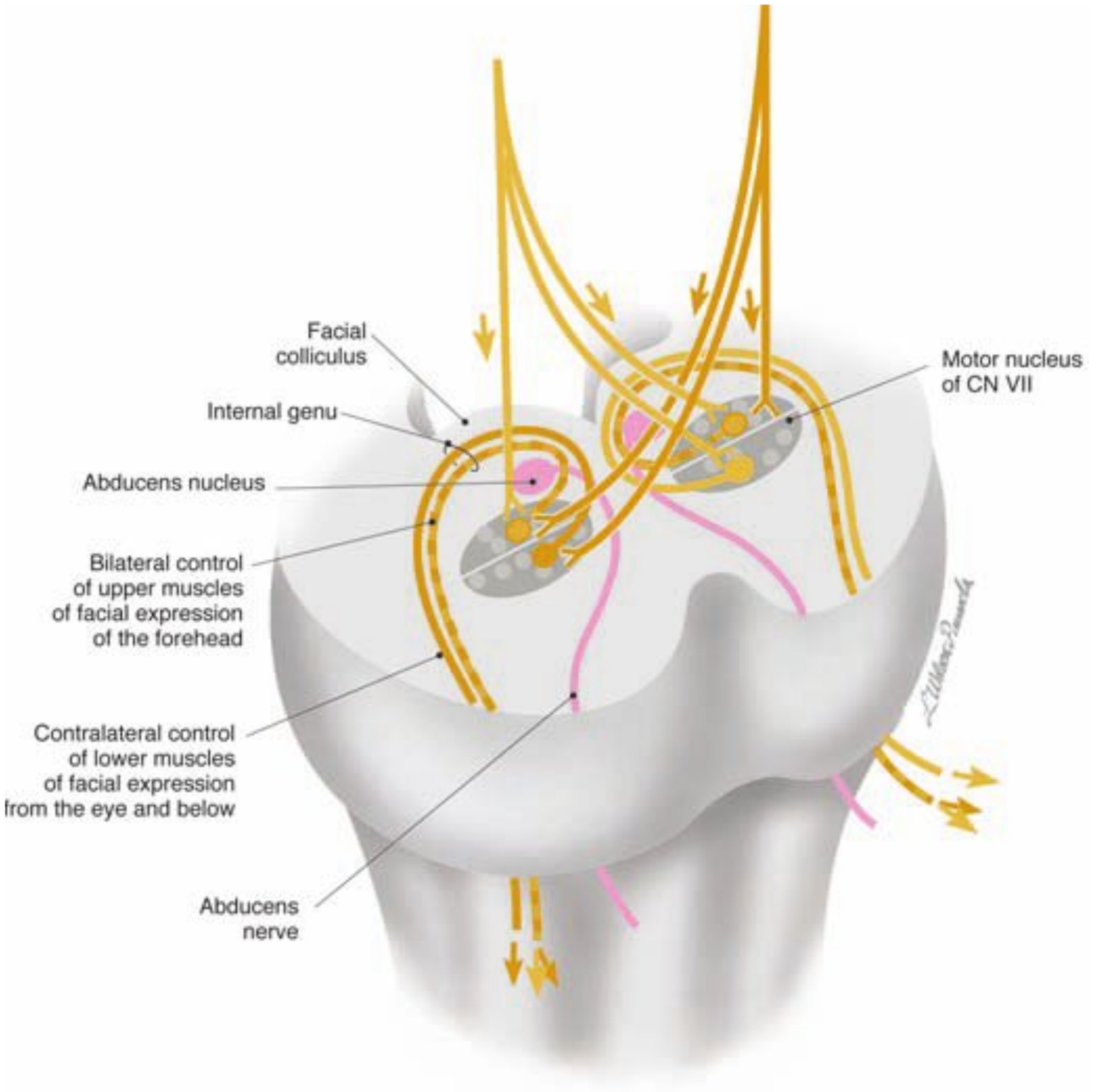

Facial colliculus
Internal genu
Abducens nucleus
Bilateral control of upper muscles of facial expression of the forehead
Contralateral control of lower muscles of facial expression from the eye and below
Abducens nerve
Motor nucleus of CN VII

FIGURE VII–8 Motor nuclei of cranial nerve VII in the pons.

Upper motor neurons that project to the parts of the nucleus innervating upper facial muscles project bilaterally, but those that project to the parts of the nucleus innervating the lower facial muscles project mainly contralaterally (Fig. VII–8).

Recent studies in rhesus monkeys suggest that neurons in the anterior cingulate gyrus may supply bilateral innervation to the facial lower motor neurons that drive frontalis and orbicularis oculi muscles (Morecraft et al., 2001, 2004). The results of a functional magnetic resonance (fMRI) study are consistent with a similar pathway in humans (Hanakawa et al., 2007). If the human is similar to the rhesus monkey, then in the case of a middle cerebral artery stroke, a bilateral cingulate projection may be able to support innervation of the upper facial muscles but not of the lower ones and that might explain sparing of frontalis and occasionally orbicularis oculi.

Branchial motor axons from the facial nucleus pass through the internal acoustic meatus to the facial canal. The nerve to stapedius is given off in the vertical part of the canal. The main trunk of the facial nerve leaves the facial canal at the stylomastoid foramen, immediately gives branches to the stylohyoid and the posterior belly

TABLE VII–2 Facial Nerve Branches to Face and Neck Muscles

Named Branches	Muscles Supplied
Nerve to stapedius	Stapedius
Nerve to stylohyoid	Stylohyoid
Nerve to posterior belly of digastric	Posterior belly of digastric
Posterior auricular nerve	Auricular branch to auricularis posterior and muscles on the cranial aspect of the auricle; occipital branch to occipital belly of occipitofrontalis
Temporal nerve	Twigs to the intrinsic muscles on lateral surface of the auricle and anterior and superior auricular muscles, anterior branches to the frontal belly of occipitofrontalis, orbicularis oculi, and corrugator supercilii
Zygomatic nerve	Orbicularis oculi, levator labii superioris, levator labii superioris alaequae nasi, zygomaticus major, levator anguli oris, risorius
Buccal nerve	Upper branches to zygomaticus major, levator labii superioris; lower deep branches to buccinator and orbicularis oris, levator anguli oris, risorius
Mandibular nerve	Mentalis, depressor labii inferioris, depressor anguli oris.
Cervical nerve	Platysma

of digastric muscles, and forms the posterior auricular nerve to the occipitalis muscle. The remaining branchial motor fibers pass anteriorly to pierce and lie within the substance of the parotid gland. At this point, the nerve divides into the temporal, zygomatic, buccal, mandibular, and cervical branches to supply the muscles of the scalp, face, and neck (Table VII–2; see also Fig. VII–7).

The branchial motor axons of the facial nerve form the efferent component of several reflex arcs: closing the eye in response to touching the cornea (corneal reflex) or to bright light (light reflex); contraction of the stapedius muscles in response to sound intensity (stapedius reflex); and sucking in response to touch sensations in the mouth (sucking reflex). As well, the facial muscles are instrumental in communicating emotional states, that is, fear, anger, surprise, disgust, sadness, and joy. The pathways for emotionally driven facial expressions are not yet known, but they probably include the limbic system and basal ganglia connections within the forebrain.

Clinical Comment

Patients with Parkinson's disease have difficulty initiating movement in the muscles of facial expression. As a result, they have a characteristic, apparently emotionless, expression as well as problems articulating speech.

PARASYMPATHETIC MOTOR (VISCERAL EFFERENT) COMPONENT

The parasympathetic component of CN VII is responsible for control of the lacrimal, submandibular, and sublingual glands and the mucous glands of the mouth, pharynx, and nose (i.e., all the major glands of the head except the integumentary glands and the parotid gland). The cell bodies (preganglionic autonomic motor neurons) are scattered in the pontine tegmentum and are collectively called the "superior salivatory nucleus." The parasympathetic motor component is illustrated in Figure VII–9.

The superior salivatory nucleus is influenced primarily by the hypothalamus, which is an important controlling and integrating center of the autonomic nervous system. Impulses from the limbic system (emotional behavior) and the olfactory

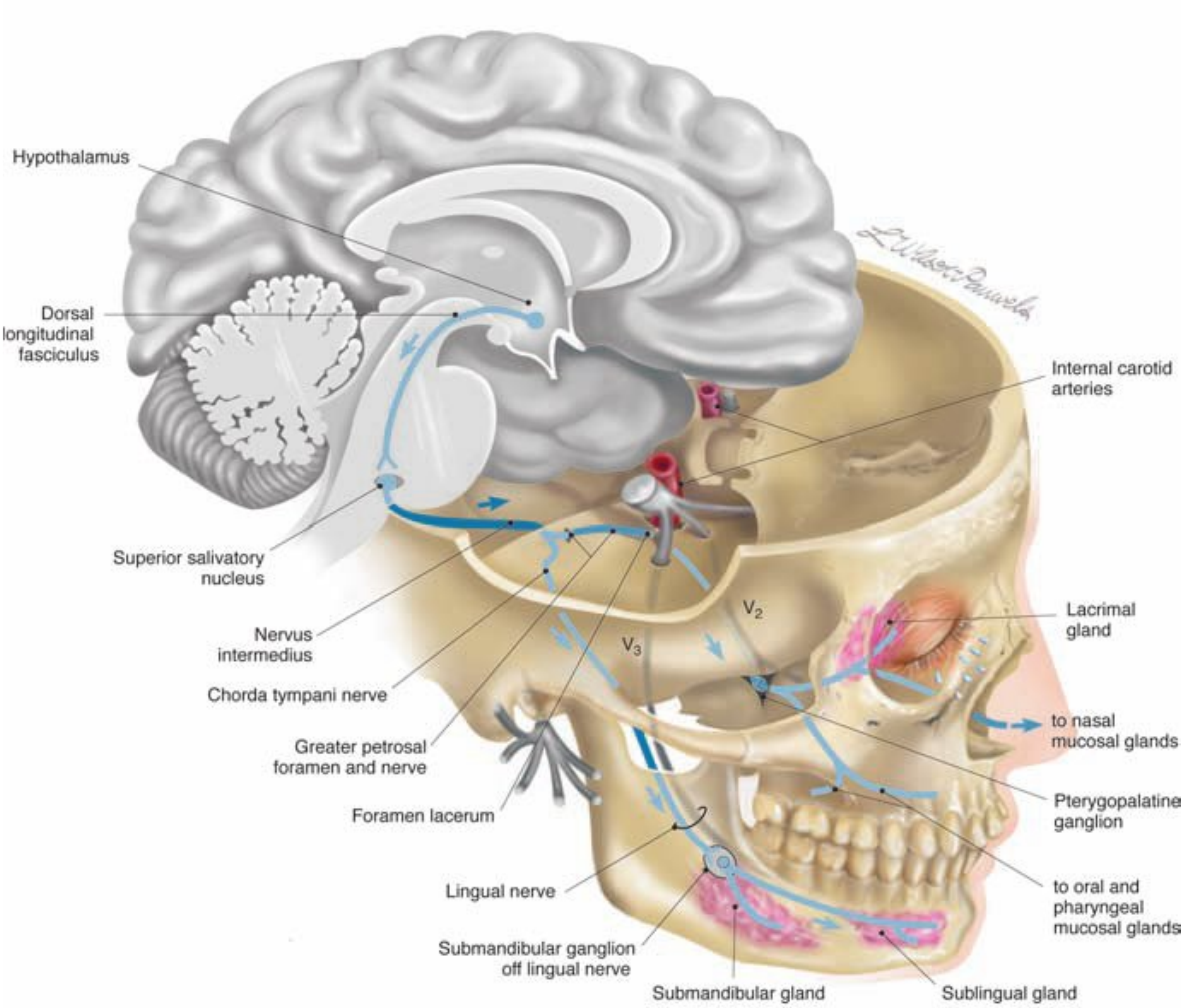


FIGURE VII–9 Parasympathetic motor component of the facial nerve.

area (special sensory area for smell) enter the hypothalamus and are relayed via the dorsal longitudinal fasciculus to the superior salivatory (lacrimal) nucleus. These pathways mediate visceral reflexes such as salivation in response to appetizing odors or weeping in response to emotional states.

The superior salivatory nucleus is also influenced by other areas of the brain. For example, when the eye is irritated, sensory fibers travel via the spinal trigeminal nucleus in the brain stem to the reticular formation, which in turn, stimulates the superior salivatory nucleus to cause secretion of the lacrimal gland. When special taste fibers in the mouth are activated, the nucleus solitarius (rostral gustatory portion) stimulates the superior salivatory nucleus to cause secretion of the submandibular and sublingual salivatory glands and the oral mucosal glands.

The efferent fibers from the superior salivatory nucleus travel in the nervus intermedius. They divide at the geniculate ganglion in the facial canal into two groups to become the greater petrosal nerve (to the lacrimal gland and to nasal, pharyngeal, and oral mucosal glands) and part of chorda tympani (to the submandibular and sublingual salivary glands and oral mucosal glands).

The greater petrosal nerve exits the petrous portion of the temporal bone via the greater petrosal foramen to enter the middle cranial fossa. It passes deep to the trigeminal ganglion to reach the foramen lacerum. It helps to think of the foramen lacerum as a short vertical chimney. The greater petrosal nerve traverses the lateral wall of the chimney to reach the pterygoid canal. Here it joins with the deep petrosal nerve (sympathetic fibers from the plexus that surrounds the internal carotid artery) to become the nerve of the pterygoid canal (Fig. VII–10). This canal is located in the base of the medial pterygoid plate of the sphenoid bone and opens into the pterygopalatine fossa where the pterygopalatine ganglion is located. Axons of parasympathetic neurons in the nerve of the pterygoid canal synapse in the parasympathetic pterygopalatine ganglion. Postganglionic fibers continue forward, some with branches of V_2, to reach the lacrimal gland and the oral, pharyngeal, and nasal mucosa where they stimulate secretion.

The chorda tympani nerve passes through the petrotympanic fissure to join the lingual branch of the mandibular nerve (V_3) after the latter has passed through the foramen ovale. These two nerve bundles travel together toward the lateral border of the floor of the oral cavity where the parasympathetic fibers of CN VII synapse in the submandibular ganglion, which is suspended from the lingual nerve. Postganglionic fibers continue to the submandibular and sublingual glands and to minor salivatory glands in the floor of the mouth where they stimulate secretion.

CASE HISTORY GUIDING QUESTIONS

1. What is Bell's palsy and what causes this disorder?
2. Why did John have trouble contracting the muscles on the right side of his face and neck?

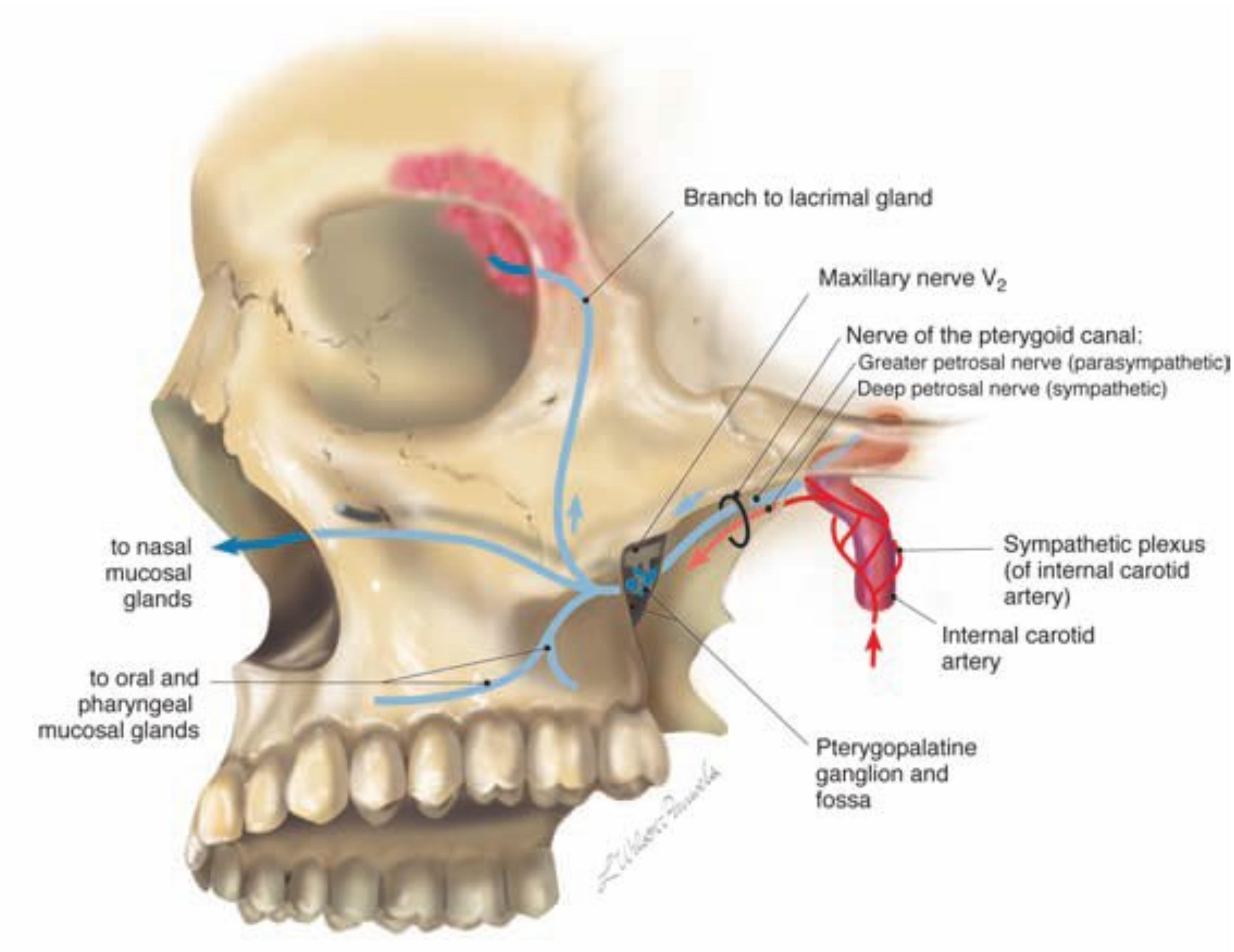

Branch to lacrimal gland
Maxillary nerve V_2
Nerve of the pterygoid canal:
Greater petrosal nerve (parasympathetic)
Deep petrosal nerve (sympathetic)
to nasal mucosal glands
Sympathetic plexus (of internal carotid artery)
Internal carotid artery
to oral and pharyngeal mucosal glands
Pterygopalatine ganglion and fossa

FIGURE VII–10 Parasympathetic motor nerve in the pterygoid canal.

3. Why was John's mouth dry?
4. Why did John perceive sounds to be louder in his right ear?
5. What is the anatomic basis for the loss of taste to the anterior two-thirds of John's tongue on the right side?
6. What is the anatomic basis of John's excessive tearing?
7. How does an understanding of John's excessive tearing problem help you to locate the site of the lesion?
8. Why was John's recovery so prolonged?
9. Why does John's right eye close every time he smiles?
10. What other lesions could affect the motor component of the facial nerve?

1. What is Bell's palsy and what causes this disorder?

Bell's palsy is paralysis of the facial nerve of unknown cause (idiopathic). It occurs when the nerve is compressed as it passes through the facial canal in the petrous portion of the temporal bone. It affects both men and women equally, and the incidence is 23 per 100,000 annually. The signs and symptoms of Bell's palsy are determined by which branches of the facial nerve are affected. Facial nerve palsy can involve the motor, secretory, and sensory components. The primary symptom in involvement of the motor branch is acute onset of unilateral weakness, reaching its maximum paralysis in 48 to 72 hours. Impairment of taste and hyperacusis (sounds seem louder than normal) can occur from involvement of the chorda tympani nerve and the nerve to stapedius, respectively. Parasympathetic nerve involvement can result in the reduction of secretory gland function. Eighty percent to 85% of patients make a full recovery within 3 months of the onset of symptoms. Recovery of some motor function in the first 5 to 7 days is the most favorable sign. A viral etiology (cause) is suspected but unproven.

2. Why did John have trouble contracting the muscles on the right side of his face and neck?

Damage to the motor pathway anywhere along its course can result in facial paralysis. In John's case, the motor axons were injured in the facial canal (Fig. VII–11) As a result, he was unable to tense the facial muscles on the right for shaving, close his right eye, or keep food in his mouth when he was eating.

3. Why was John's mouth dry?

The facial nerve carries parasympathetic secretomotor fibers to the submandibular and sublingual glands as well as to the mucosa of the mouth. Because John's mouth was dry, we know that the lesion to the facial nerve must have occurred before the parasympathetic component left the facial nerve as part of the chorda tympani. This would mean that, as well as branchial motor loss, the secretomotor action of the facial nerve would be lost and the production of saliva decreased (see Fig. VII–11).

4. Why did John perceive sounds to be louder in his right ear?

The sound transducing mechanism in the inner ear can be damaged by loud sounds. The stapedial reflex helps to protect the inner ear by limiting the amount of energy transmitted through the chain of small bones in the middle ear. When sound entering the ear is above 77 decibels, and just before the onset of speaking, the stapedial muscle contracts, thereby limiting the movement of the stapes. In John's case, the lesion was above the origin of the nerve to stapedius; therefore, the stapedial reflex on the right side was not working and more sound energy was delivered to his right inner ear (see Fig. VII–11).

5. What is the anatomic basis for the loss of taste to the anterior two-thirds of John's tongue on the right side?

Loss of taste to the anterior two-thirds of the tongue on the right side can result from a lesion of the right facial nerve anywhere along its course to the point at which the chorda tympani nerve leaves the main trunk in the facial canal or to the chorda tympani itself. To locate the specific site of the lesion, other deficits as well as the loss of taste must be identified. In John's case, he has paralysis of all the muscles supplied by the facial nerve as well as the loss of secretion of saliva, loss of taste to the anterior

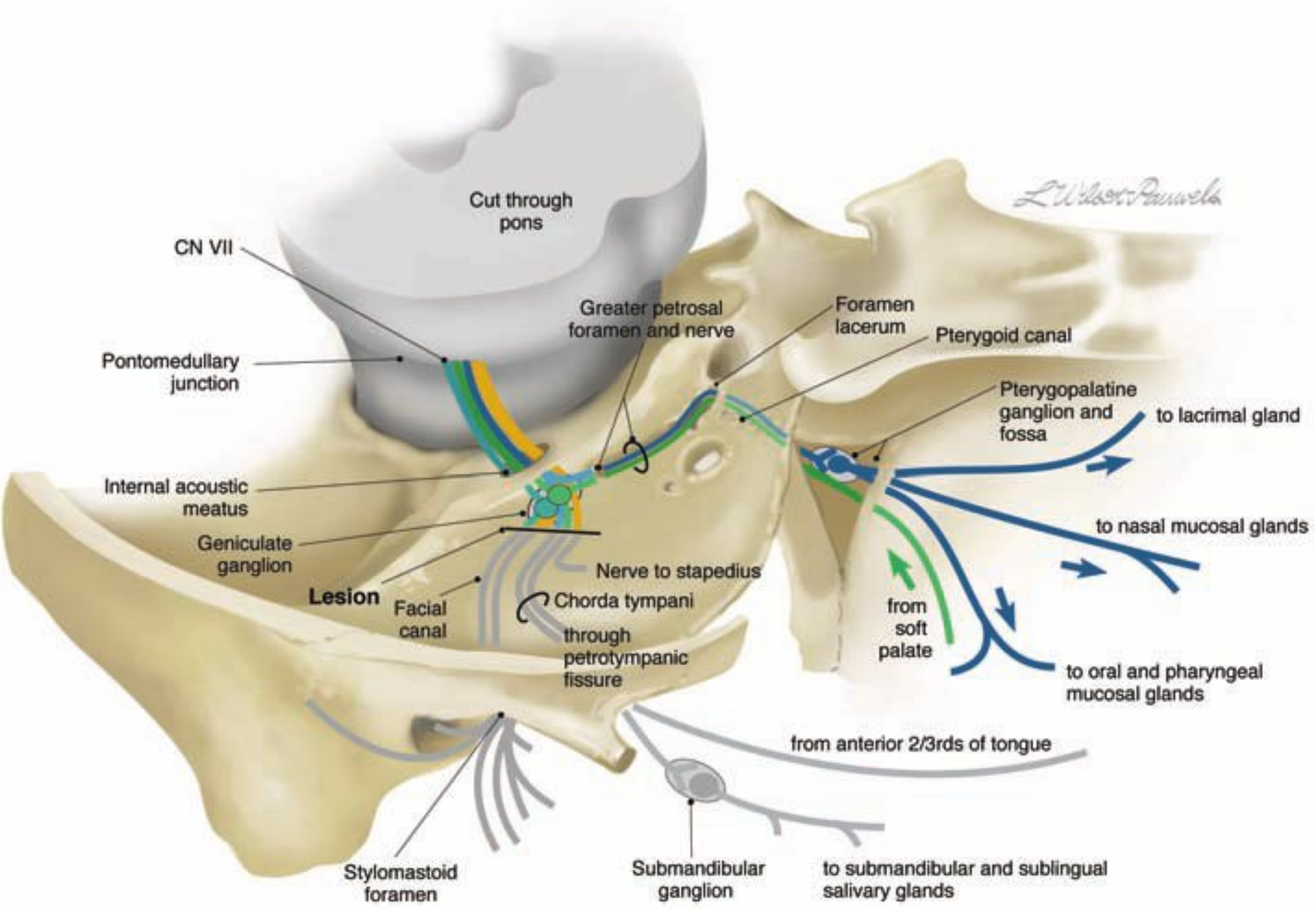


FIGURE VII–11 Facial nerve lesion distal to the geniculate ganglion.

two-thirds of the tongue, and loss of the stapedial reflex. The lesion, therefore, must be in the facial canal proximal to the branch to stapedius (see Fig. VII–11).

6. What is the anatomic basis of John's excessive tearing?

Normally, only enough tears are produced to replace fluid lost by evaporation from the surface of the eye. Rhythmic blinking moves the tears from the upper lateral aspect of the orbit where the lacrimal gland is located to the medial corner where the collecting apparatus is located. This constant washing of the cornea keeps it moist. Because of the paralysis of his orbicularis oculi muscle, John cannot blink his right eye, and his right cornea dries out. This has two consequences: (1) The dryness and irritation of his cornea, signaled by cranial nerve V_1 (part of the afferent limb of the blink reflex), stimulate the superior salivatory nucleus, which in turn, sends secretomotor signals via the greater petrosal nerve (CN VII) to the lacrimal gland to produce and release an increased volume of tears. (2) The excess tears cannot be moved to the collecting ducts in the medial aspect of the eye, so they collect in his right conjunctival sac and spill over onto his cheek.

7. How does an understanding of John's excessive tearing problem help you to locate the site of the lesion?

The presence of tearing is helpful in the anatomic localization of the lesion. The parasympathetic fibers branch from CN VII at the geniculate ganglion and become the greater petrosal nerve. As a result, if a lesion is proximal to the geniculate ganglion, all modalities of CN VII, including tearing, will be lost. If the lesion is distal to the geniculate ganglion, tearing is not affected. Because John is able to produce tears in his right eye, we know that his lesion must be located in the facial nerve distal to the geniculate ganglion (see Fig. VII–11).

8. Why was John's recovery so prolonged?

Unfortunately, John suffered from axonal damage to his facial nerve. Typically, in Bell's palsy, there is no axonal damage. However, sometimes axonal loss does occur and there is electromyographic evidence of muscle degeneration in the face. In this case, recovery is prolonged because it is dependent on regeneration of the nerve, which may take years.

9. Why does John's right eye close every time he smiles?

After axonal damage of the right facial nerve, there may be regeneration with the risk of misdirection of the regenerating fibers. Aberrant reinnervation of facial muscles results in abnormal synkinesis (motor activity). For example, if some fibers from the motor neurons that initiate smiling regenerate and join with neurons innervating the orbicularis oculi, the result could be closure of the eye when the patient smiles spontaneously. Conversely, if some of the motor fibers that innervate orbicularis oris regenerate to join neurons that activate the labial muscles, the result could be a twitch of the mouth when the patient blinks.

In rare cases, the regenerating fibers originally supplying the submandibular and sublingual salivary glands aberrantly innervate the lacrimal gland via the greater petrosal nerve. This results in inappropriate unilateral lacrimation with eating. This condition is called "crocodile tears."

10. What other lesions could affect the motor component of the facial nerve?

A facial palsy can result from a lesion anywhere along the motor pathway from the cortex to the muscles. Lesions are classified as upper motor neuron lesions or lower motor neuron lesions, depending on their location.

Upper Motor Neuron Lesions

Cortical tumors, infarcts, and abscesses affecting upper motor neuron cell bodies in the motor cortex, or their axons that project to the facial nucleus, result in loss of voluntary control of mainly the lower muscles of facial expression contralateral to the lesion (Fig. VII–12A). The frontalis muscle continues to function because the part of the facial nucleus that innervates its fibers still receives input from the ipsilateral hemisphere (see Fig. VII–8, expanded view of the nucleus).

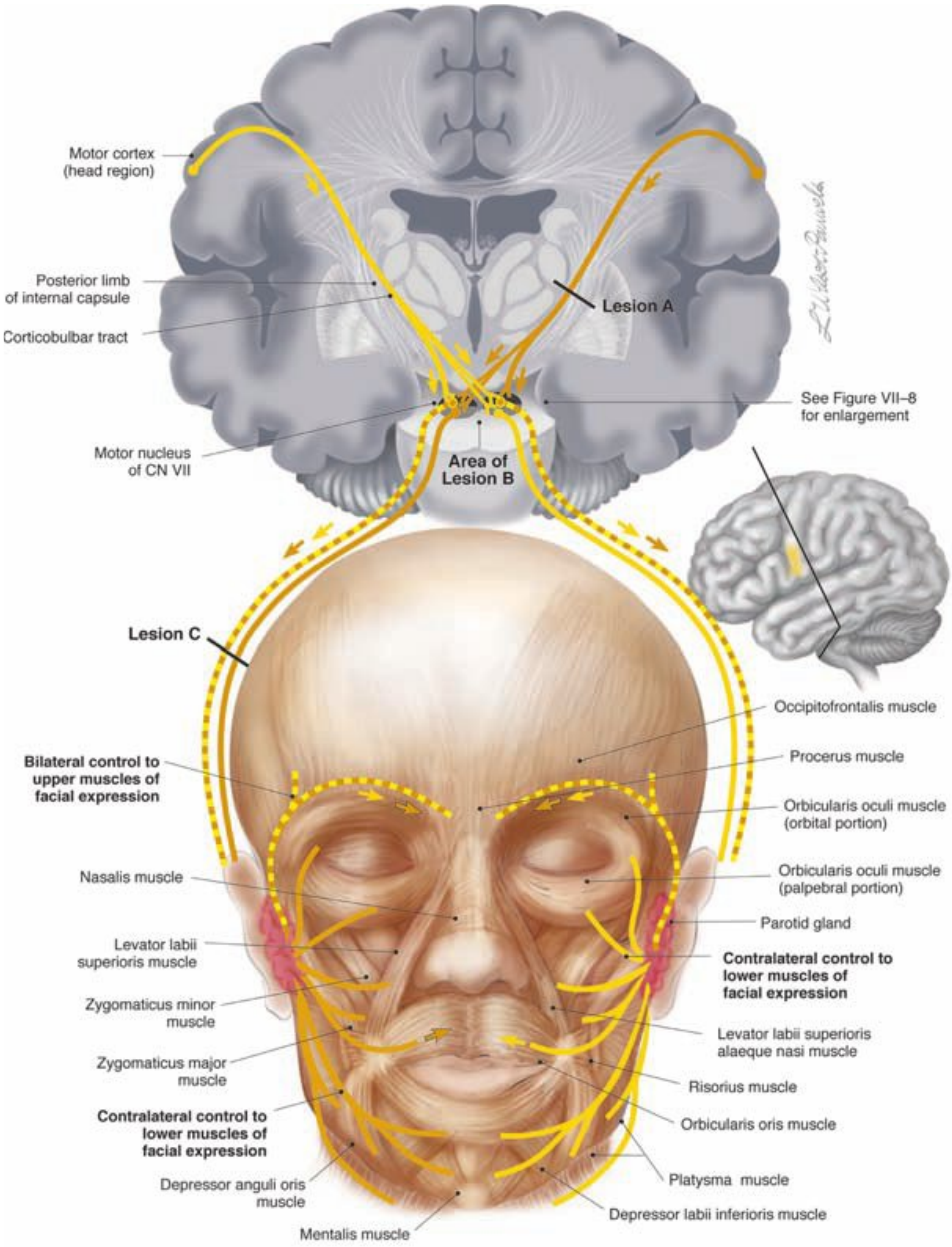

Motor cortex (head region)
Posterior limb of internal capsule
Corticobulbar tract
Lesion A
See Figure VII–8 for enlargement
Motor nucleus of CN VII
Area of Lesion B
Lesion C
Occipitofrontalis muscle
Procerus muscle
Bilateral control to upper muscles of facial expression
Orbicularis oculi muscle (orbital portion)
Orbicularis oculi muscle (palpebral portion)
Nasalis muscle
Parotid gland
Levator labii superioris muscle
Contralateral control to lower muscles of facial expression
Zygomaticus minor muscle
Levator labii superioris alaeque nasi muscle
Zygomaticus major muscle
Risorius muscle
Contralateral control to lower muscles of facial expression
Orbicularis oris muscle
Platysma muscle
Depressor anguli oris muscle
Depressor labii inferioris muscle
Mentalis muscle

FIGURE VII–12 Innervation of muscles of facial expression (not all muscles are illustrated). Both cerebral hemispheres innervate the lower motor neurons that drive the frontalis muscle (dotted lines). Only the contralateral cortex innervates the lower motor neurons that drive the remaining muscles of facial expression (solid lines).

The most common upper motor neuron lesion that involves CN VII is a vascular infarct or stroke that damages neurons in the cortex or, more commonly, their axons in the internal capsule. These axons may also be affected anywhere along their course.

Lower Motor Neuron Lesions

Lesions resulting from damage to the facial nucleus or its axons anywhere along the course of the nerve after it leaves the nucleus (see Fig. VII–12B and C) are commonly called “lower motor neuron lesions.”

Lesions in the pons (Fig. VII–13A; see also Fig. VII–12B) that involve the nucleus of CN VII are commonly due to an infarct involving the pontine branches of the basilar artery. This results in a complete paralysis of the facial nerve ipsilaterally, combined with motor weakness of the limbs on the contralateral side of the body due to damage of the descending corticospinal fibers from the motor cortex before they cross over in the medulla. Such lesions often include paralysis of the lateral rectus muscle as well, because the abducens nucleus is in close proximity to the facial

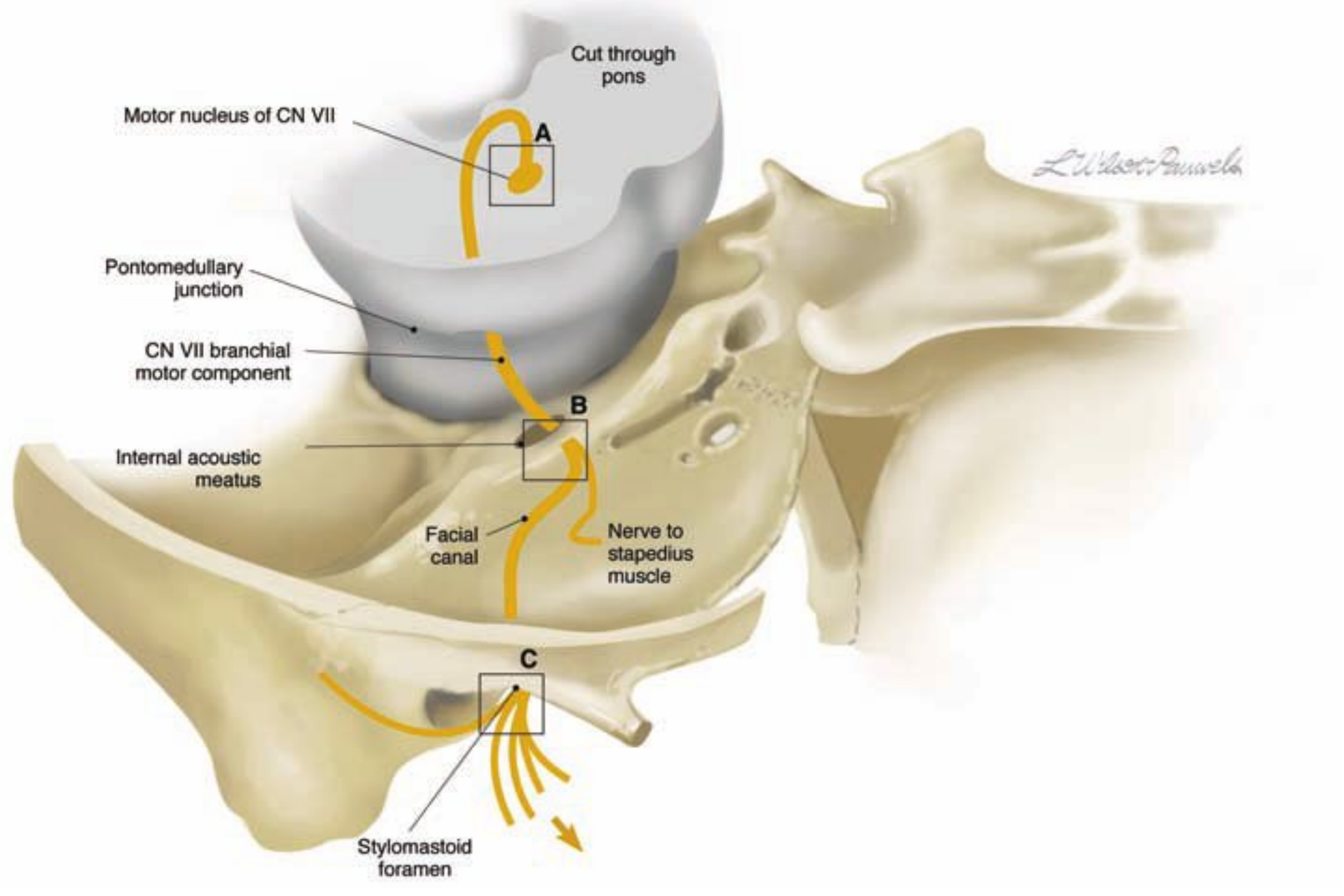


FIGURE VII–13 Lower Motor Neuron Lesions (LMNLs) affecting the branchial motor axons in **A**, pons, **B**, internal acoustic meatus, and **C**, styloid mastoid foramen (brainstem is elevated).

nucleus (see Fig. VII–8). Pontine tumors may also disrupt the facial nucleus and nearby structures.

As the nerve exits the brain stem, it may be damaged by acoustic neuromas (see Chapter VIII), meningiomas, and meningitis. Typically, CNs VI, VIII, and sometimes V are also involved at this site. After entering the facial canal (see Fig. VII–13B), the nerve is susceptible to fractures of the base of the skull, spread of infection from the middle ear, herpes zoster infections, and, as in John's case, idiopathic (unknown cause) Bell's palsy. All the muscles supplied by the nerve are paralyzed ipsilateral to the lesion. All actions of the facial muscles, whether motivated by voluntary, reflex, or emotional input, are affected and there is atrophy of the facial muscles. In an infant the mastoid process is not well developed and the facial nerve is very close to the surface where it emerges from the stylomastoid foramen. The use of forceps during deliver can, therefore, compromise the facial nerve (see Fig. VII–13C).

When examining a patient with facial paralysis, it is important to determine whether the paralysis is due to an upper motor neuron lesion or a lower motor neuron lesion. Asking the patient to raise his or her eyebrows can answer this question. If he or she has an upper motor neuron lesion, he or she will be able to raise both eyebrows (Fig. VII–14A) because the lower motor neurons that drive the frontalis muscle receive signals from both cerebral hemispheres (see Fig. VII–14B). Therefore, loss of signals from the affected hemisphere will not result in paralysis of this muscle. If the patient is unable to raise his or her eyebrow on the affected side, he or she has a lower motor neuron lesion (see Fig. VII–14C).

CLINICAL TESTING

When testing CN VII, it is important to examine the five main functions (see also "Cranial Nerves Examination" the web site).

1. Function of the Muscles of Facial Expression

When examining the muscles of facial expression, the examiner should first observe the patient's facial movements when he or she is speaking. Next, the examiner should ask the patient to raise his or her eyebrows to assess the action of the frontalis muscle. Symmetrical furrowing of the forehead indicates normal function (Fig. VII–15A). Orbicularis oculi muscle is tested by asking the patient to close his or her eyes as tightly as possible. This should result in burrowing of the eyelashes, and the examiner should be unable to open the patient's eyes when the patient resists (see Fig. VII–15B). The buccinator and orbicularis oris muscles are tested by asking the patient to press the lips firmly together. If there is full strength, the examiner should be unable to separate the patient's lips (see Fig. VII–15C). The platysma muscle can be tested by asking the patient to clench the jaw, and the examiner should see the tightening of the muscle as it extends from the body of the mandible downward over the clavicle onto the anterior thoracic wall.

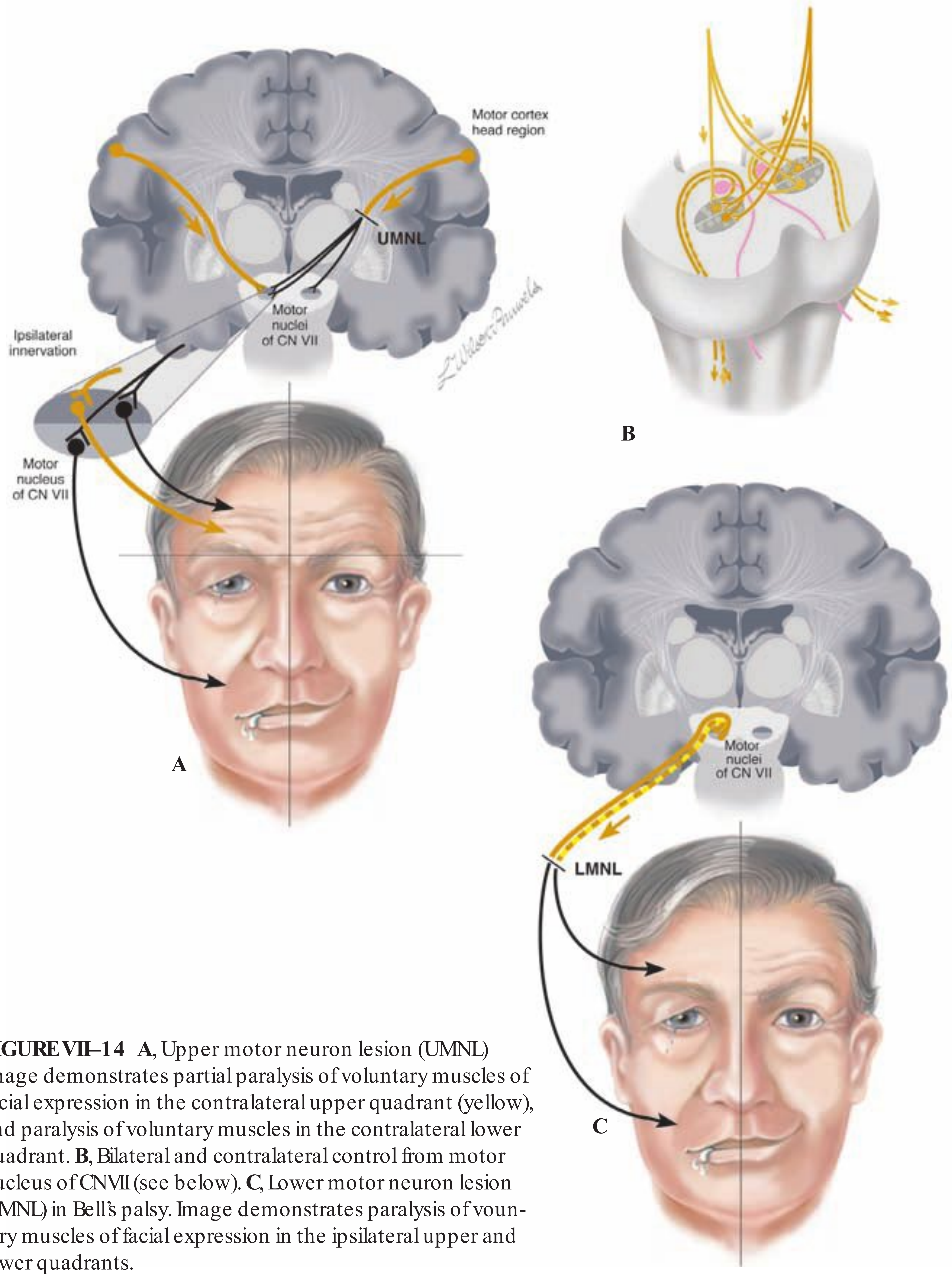


FIGURE VII–14 **A**, Upper motor neuron lesion (UMNL) Image demonstrates partial paralysis of voluntary muscles of facial expression in the contralateral upper quadrant (yellow), and paralysis of voluntary muscles in the contralateral lower quadrant. **B**, Bilateral and contralateral control from motor nucleus of CN VII (see below). **C**, Lower motor neuron lesion (LMNL) in Bell's palsy. Image demonstrates paralysis of vountary muscles of facial expression in the ipsilateral upper and lower quadrants.

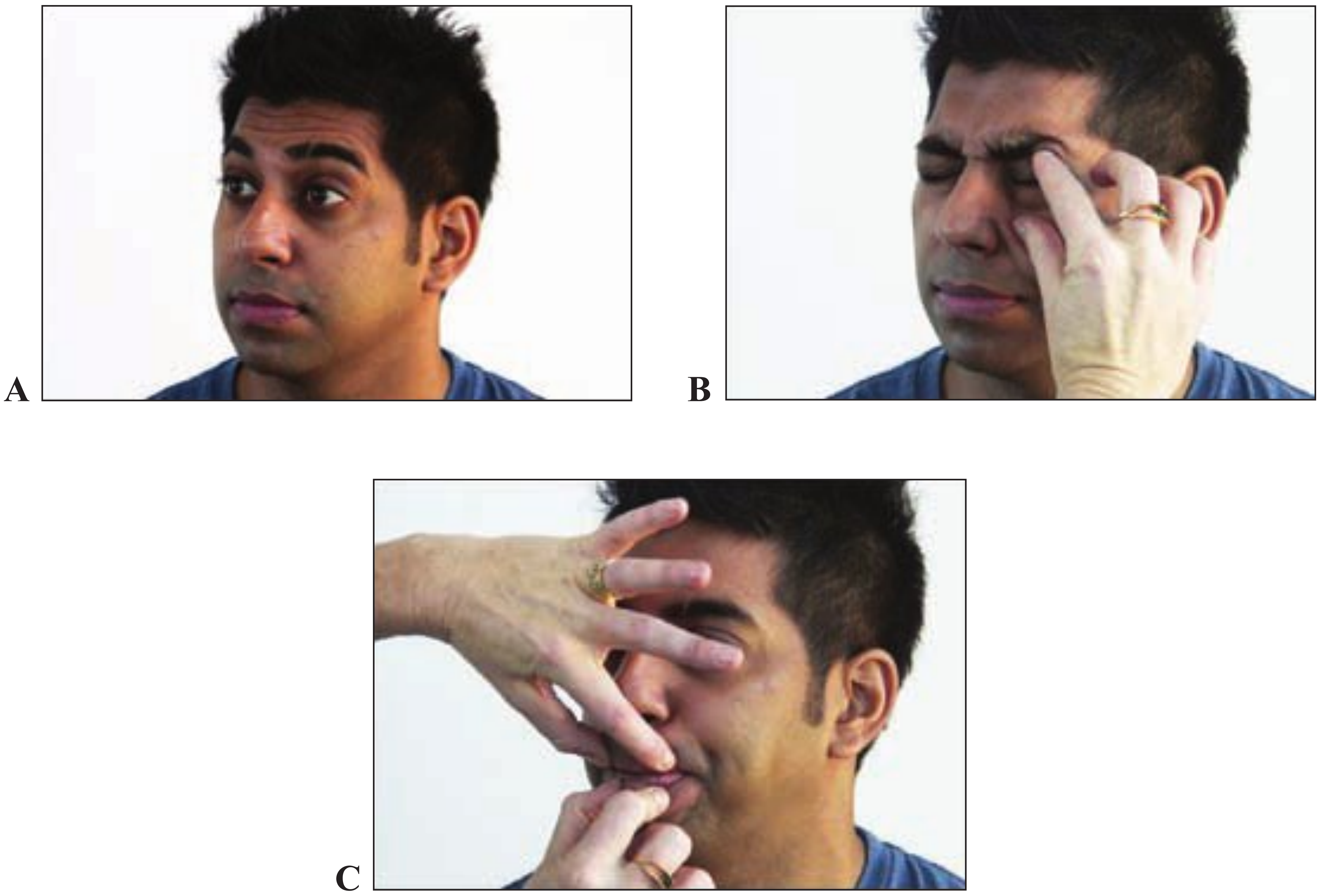


FIGURE VII–15 Clinical tests for the muscles of facial expression. **A**, Frontalis muscle. **B**, Orbicularis oculi muscle. **C**, Orbicularis oris muscle.

2. Taste from the Taste Buds

CN VII includes special sensory axons that carry taste sensation from the anterior two-thirds of the tongue in the chorda tympani to the nucleus solitarius via the nervus intermedius. This modality is assessed using a stick with a piece of cotton moistened in a sugary or salty solution. The patient is asked to protrude his or her tongue and the examiner touches the tongue on one side with the solution (Fig. VII–16A). Before the patient returns his or her tongue into the mouth, he or she is asked to report what he or she tasted by pointing to the appropriate text on a sign, and then the examiner applies the solution to the other side of the tongue and asks the patient if there is any difference between the two sides (see Fig. VII–16). The patient must rinse the mouth out with water before repeating the test with the next solution.

3. Somatic Sensation from the External Ear

A general sensory branch of CN VII provides a variable contribution to CNs V and X in innervating the concha, part of the external acoustic meatus and the external (lateral) surface of the tympanic membrane. Although sensation in this area can be tested by touching the skin with a sharp object for pain sensation and with cotton wool for light touch, abnormalities of sensation are rarely detected when this branch is involved because of overlap from other nerves that supply this small region.

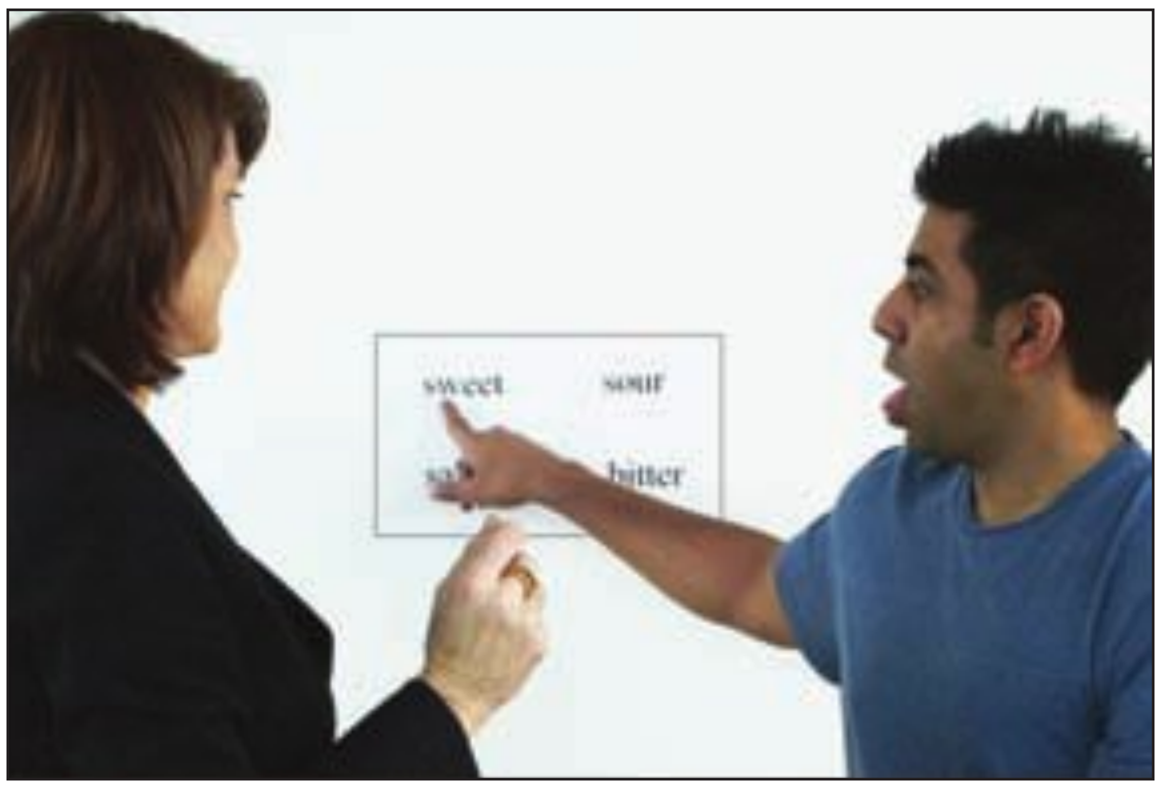


FIGURE VII–16 Clinical testing for taste. Four basic modalities (sweet, sour, bitter, and salty) are tested on each side of the tongue. After the examiner holds the tongue and touches one side of it with a cotton swab soaked in a test solution, the patient points to the taste he perceives on the board.

4. The Function of the Stapedius Muscle

The stapedius muscle is responsible for dampening the oscillations of the ossicles of the middle ear. This muscle contracts reflexively to loud noises and at the onset of speech. Therefore, an individual who has damage to this branch of the nerve will hear noises louder on the affected side. This is referred to as "hyperacusis." Hyperacusis can be assessed by standing behind the patient and suddenly clapping your hands together beside one ear and then the other ear and asking the patient if there is any difference in loudness between the two sides.

5. Secretomotor Innervation of the Lacrimal and Salivary Glands

The parasympathetic efferent fibers from the superior salivatory (lacrimal) nucleus travel in the nervus intermedius and divide into two nerves within the facial canal (see Fig. VII–9). Tearing, which is a function of the lacrimal gland, can be tested using Schirmer's test. One end of a piece of filter paper 5 mm wide and 25 mm long is inserted into the lower conjunctival sac, while the other end hangs over the edge of the lower lid. In a normal individual, the filter paper will absorb tears from the conjunctival sac and, after 5 minutes, have a moistened area that extends for approximately 15 mm along the filter paper. A moistened area of less than 10 mm is suggestive of hypolacrimia (underproduction of tears). For practical purposes, this test is not commonly done. Direct questioning of the patient about the presence of dry eyes is adequate.

Although salivatory function cannot be tested easily at the bedside, it is still possible to assess it by asking whether the patient has a dry mouth and whether water is required to help with swallowing a meal.

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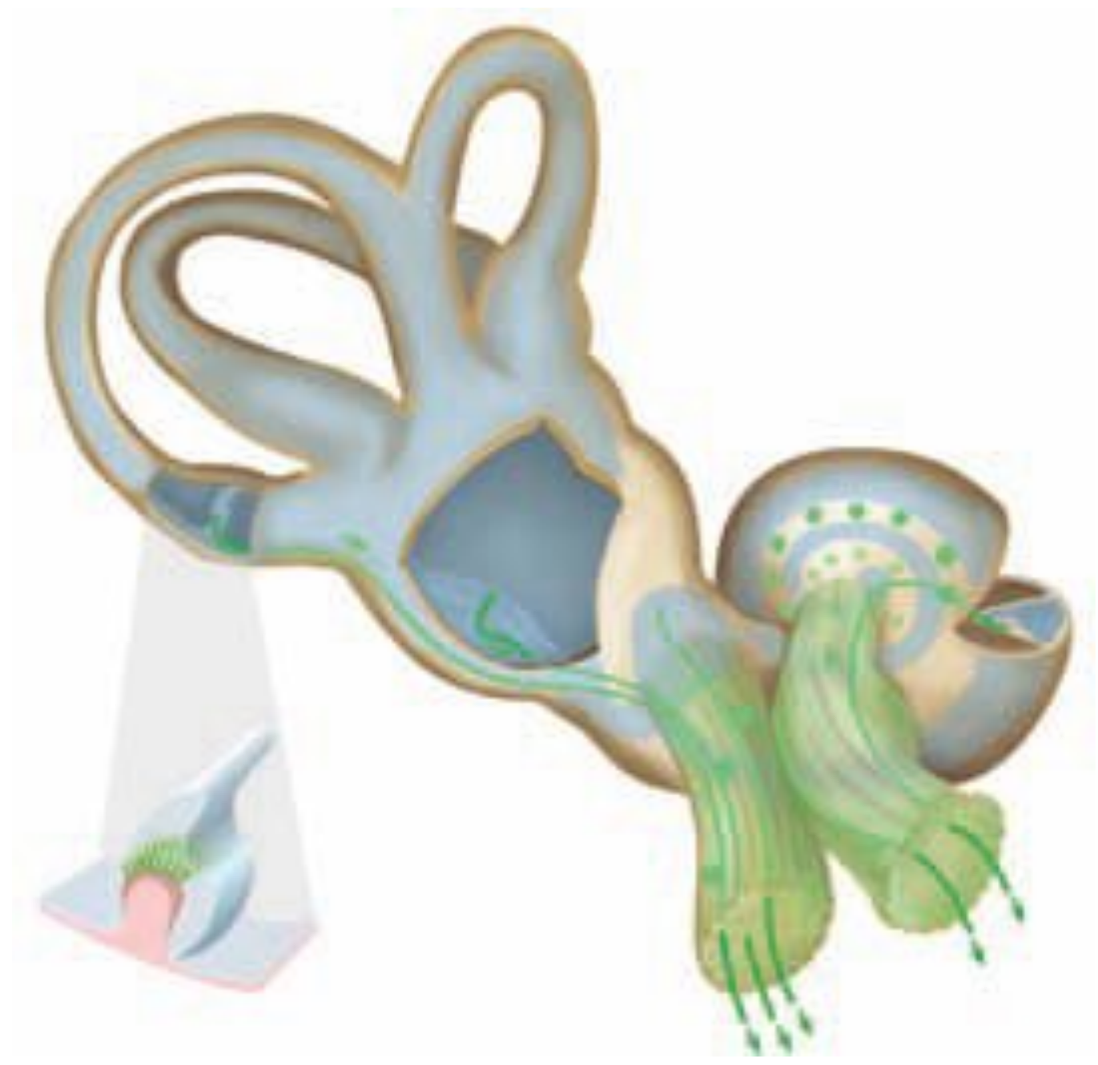


VIII

Vestibulocochlear Nerve

CASE HISTORY

Paul, a 34-year-old sports reporter, noticed that he was having difficulty hearing in his right ear while talking on the telephone. Occasionally, he also heard ringing in his right ear, which had become progressively worse over the last couple of months. Paul racewalks in his spare time and lately has felt that his balance was off. One Sunday afternoon while on a training walk, Paul experienced a wave of vertigo (dizziness), lost his balance, and hit his head on a lamp post. He received a nasty cut to his scalp, so he went to the emergency department to get it sutured. He explained to the doctor that he was having some problems with his balance. When the physician examined Paul, she observed that he had abnormal eye movements. Although his eyes were able to move fully in all directions, his eyes alternated between a slow movement in one direction and a quick, corrective movement back to the primary position (horizontal nystagmus). See "Clinical Testing" in this chapter and in Chapters III and 13. His corneal reflexes, facial sensation, and facial movements were normal. When the physician examined Paul at the bedside, she found a marked reduction of hearing in his right ear. When a Weber's test was performed (see "Clinical Testing"), Paul heard the sound louder in his left ear. The remainder of his cranial nerves (CNs) were functioning normally, but a full neurologic examination revealed that Paul had signs of cerebellar abnormalities. A magnetic resonance imaging (MRI) scan of his head was performed, and he was found to have a large tumor of his right CN VIII, which was pressing on his pons and cerebellum.

ANATOMY OF THE VESTIBULOCOCHLEAR NERVE

The vestibulocochlear nerve carries two types of special sensation: vestibular (balance) and auditory (hearing) (Table VIII–1 and Fig. VIII–1). The sensory receptors, called "hair cells," are situated in specialized areas on the inner walls of the membranous labyrinth. The membranous labyrinth is an intricate tubular structure filled with fluid (endolymph; see Figure VIII–1 and VIII–2 blue areas) lying inside

TABLE VIII–1 Components, Cells of Origin and Function of the Vestibulocochlear Nerve (CN VIII)

Component	Nucleus	Ganglion and Cell of Origin	Function
Special sensory (afferent)	Vestibular nucleus	Vestibular (Scarpa's) ganglion Vestibular hair cells	For balance
Special sensory (afferent)	Cochlear nucleus	Spiral ganglion Cochlear hair cells	For hearing

a complex of interconnected tunnels within the petrous temporal bone, called the "bony labyrinth." The epithelial cells of the lining of the bony labyrinth secrete a second fluid (perilymph), which surrounds the membranous labyrinth. Each membranous labyrinth is composed of a spiral cochlear division and a larger vestibular division that includes the vestibule (saccule and utricle) and three semicircular canals oriented approximately at right angles to each other.

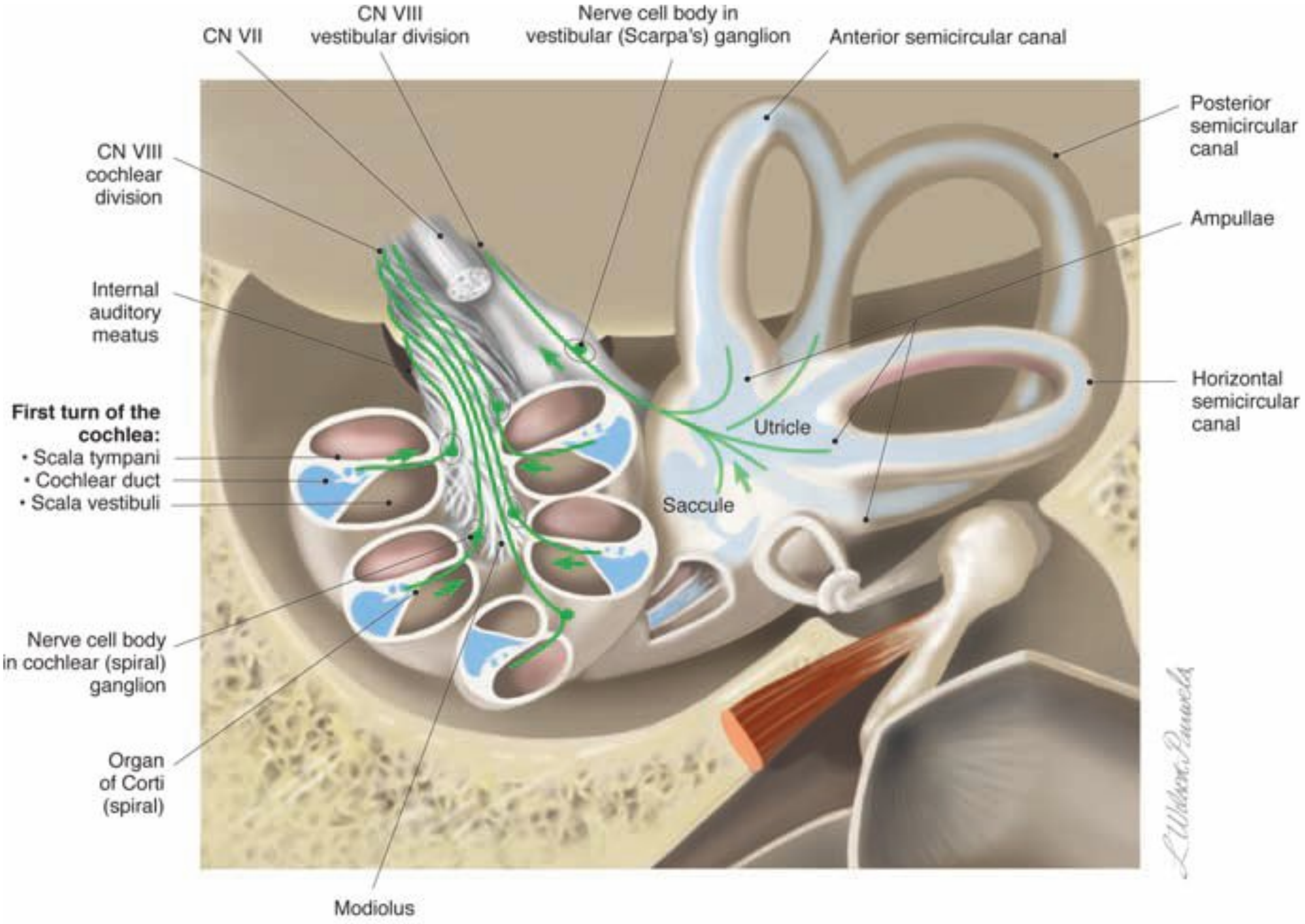

CN VII
CN VIII vestibular division
Nerve cell body in vestibular (Scarpa's) ganglion
Anterior semicircular canal
Posterior semicircular canal
CN VIII cochlear division
Ampullae
Internal auditory meatus
Horizontal semicircular canal
First turn of the cochlea:
• Scala tympani
• Cochlear duct
• Scala vestibuli
Utricle
Saccule
Nerve cell body in cochlear (spiral) ganglion
Organ of Corti (spiral)
Modiolus

FIGURE VIII–1 Overview of the vestibulocochlear nerve, cranial nerve VIII. The membranous labyrinth (blue) contains endolymph.

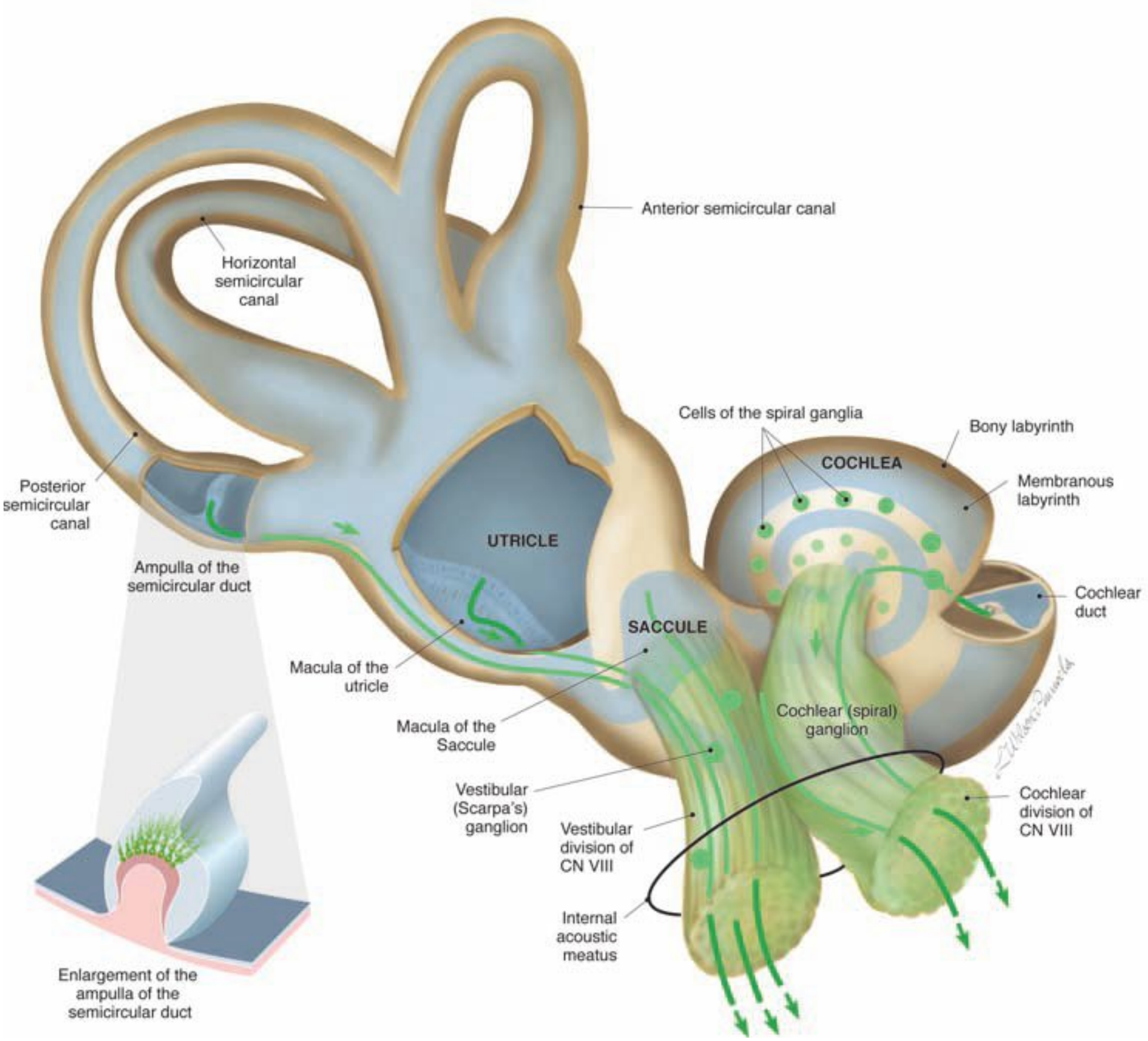

Anterior semicircular canal
Horizontal semicircular canal
Cells of the spiral ganglia
Bony labyrinth
COCHLEA
Membranous labyrinth
Posterior semicircular canal
UTRICLE
Ampulla of the semicircular duct
Cochlear duct
SACCULE
Macula of the utricle
Macula of the Saccule
Cochlear (spiral) ganglion
Vestibular (Scarpa's) ganglion
Vestibular division of CN VIII
Cochlear division of CN VIII
Internal acoustic meatus
Enlargement of the ampulla of the semicircular duct

FIGURE VIII–2 Bony and membranous labyrinths of the vestibulocochlear nerve. cranial nerve VIII (removed from the surrounding petrous temporal bone).

The cell bodies of the primary sensory neurons are located in the vestibular (Scarpa's) and spiral ganglia. These neurons are unipolar and bipolar in shape. Their peripheral (dendritic) processes extend for only a short distance from the cell bodies to the bases of the hair cells. The central (axonal) processes of these neurons form CN VIII, which travels through the internal auditory meatus in company with CN VII, and enters the posterior cranial fossa. The nerve joins the brain stem at the pontomedullary junction to terminate within the vestibular and cochlear nuclei.

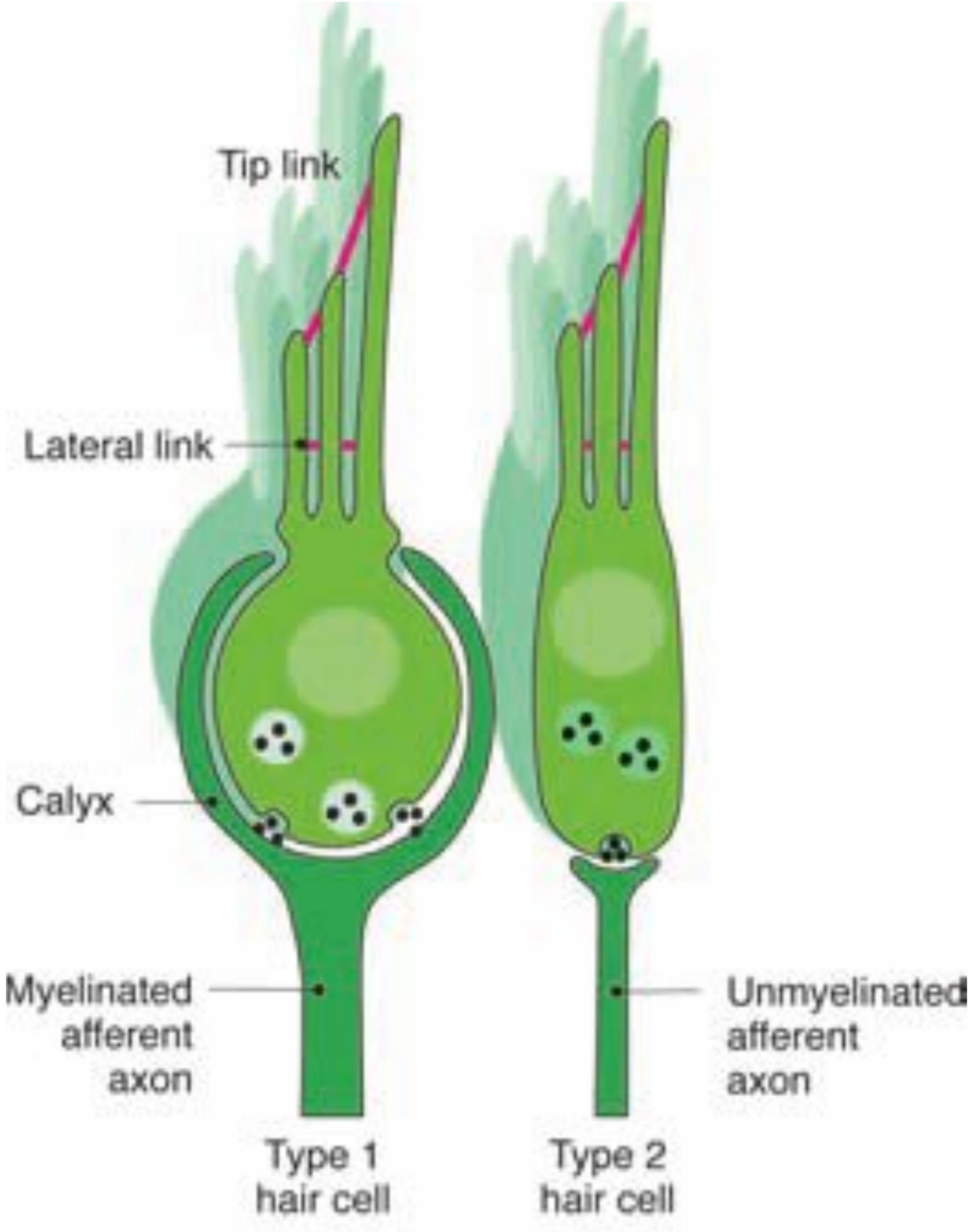


FIGURE VIII–3 Type 1 and type 2 hair cells in vestibular apparatus.

HAIR CELLS

Hair cells are specialized neurons that transduce mechanical energy into electrical signals in both the vestibular and the cochlear components of CN VIII. All hair cells have a specialized apical surface called the "cuticular plate" that is joined to surrounding cells by tight junctions and that supports a number of projecting stereocilia (Figs. VIII–3 to VIII–5). The stereocilia are of different heights and are highly ordered in rows that proceed from shortest to longest across the top of the cell. The cilia are joined along their sides by proteinaceous strands called "lateral links" that keep the cilia in a coherent bundle so that they move in unison (see Fig. VIII–3). In addition, shorter cilia are joined at their tips to the sides of longer cilia in the next row by strands called "tip links." Mechanically gated ion channels are located at the site of attachment of the tip links (see Fig. VIII–4).

A small percentage of the ion channels are open under "resting" conditions. Because the inside of the cell is negatively charged compared with the outside, positive ions move through the open channels into the cell down the electrical gradient. Potassium (K^+) is abundant in endolymph and is therefore the major ion that enters the hair cells through the open ion channels. The consequent depolarization of the membrane leads to release of neurotransmitter (glutamate) at the base of the hair cell in specialized areas called "ribbon synapses". The neurotransmitter activates nearby sensory dendrites and causes a train of impulses to be transmitted along the nerves to the brain.

When mechanical disturbances cause the cilia to sway back and forth, they move in a single plane toward and away from the tallest cilium. As they move toward the tallest cilium, the tip links stretch and put tension on the mechanically gated ion channels, causing more of them to open. As a result, more potassium moves into

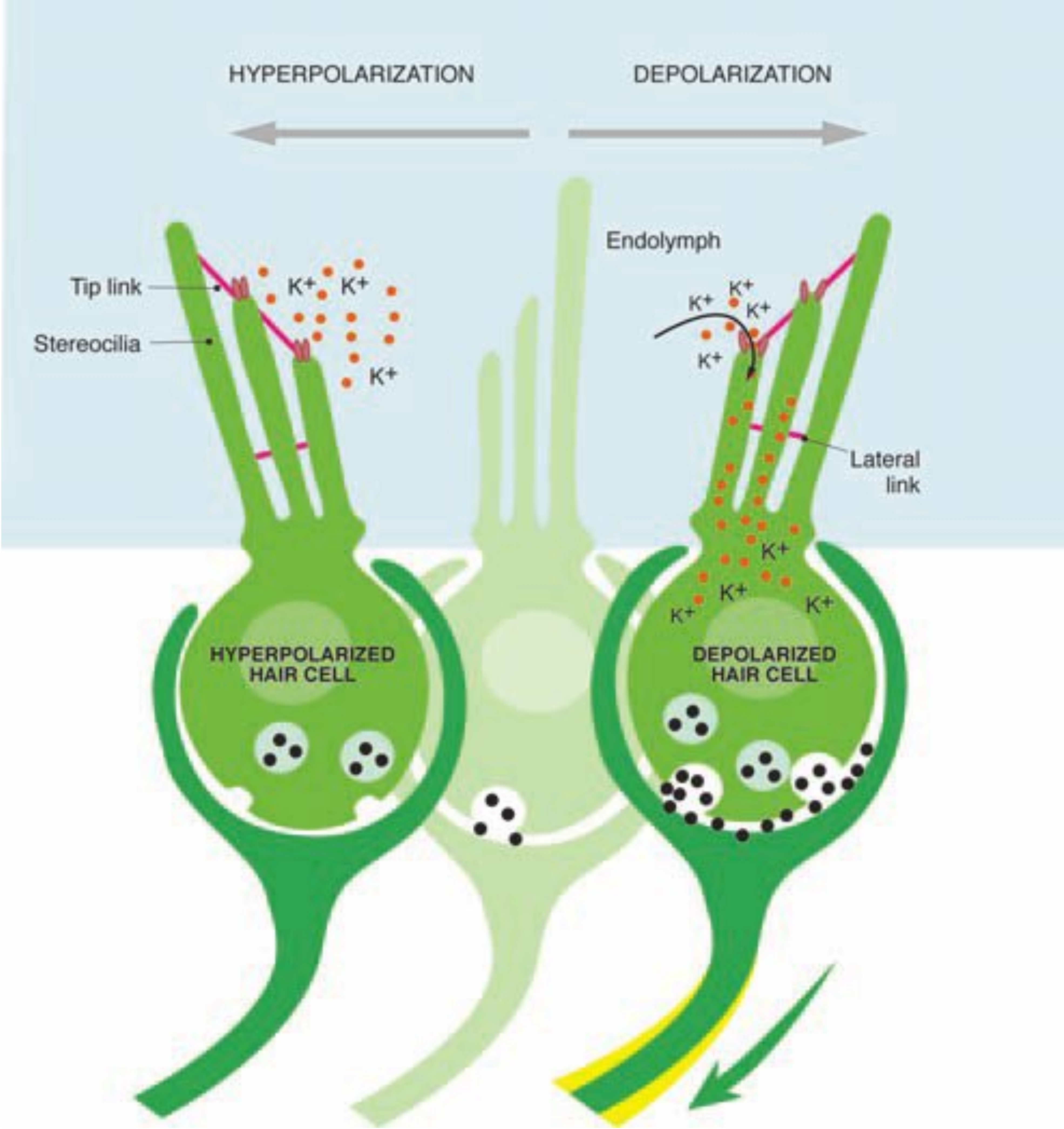
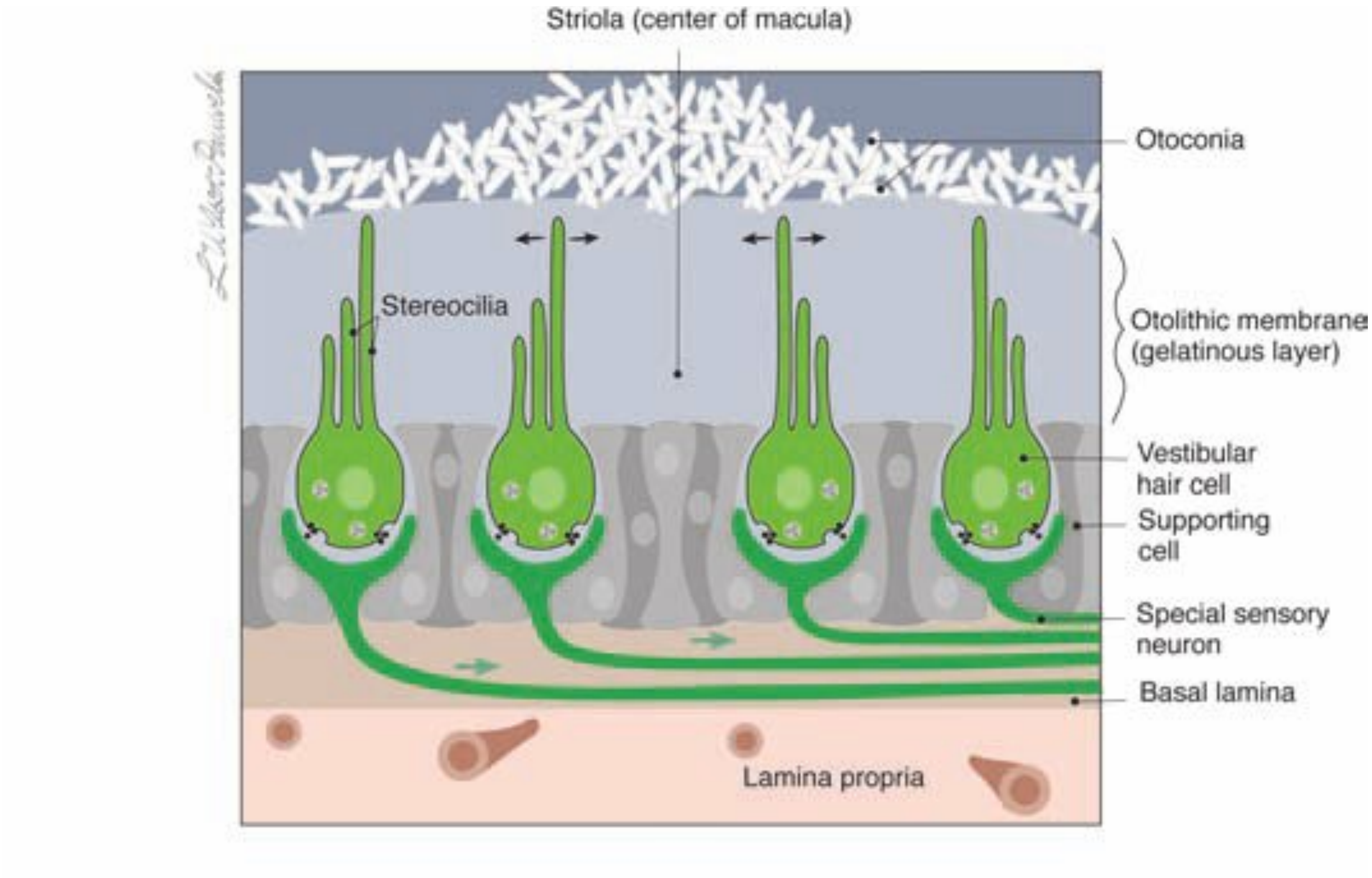
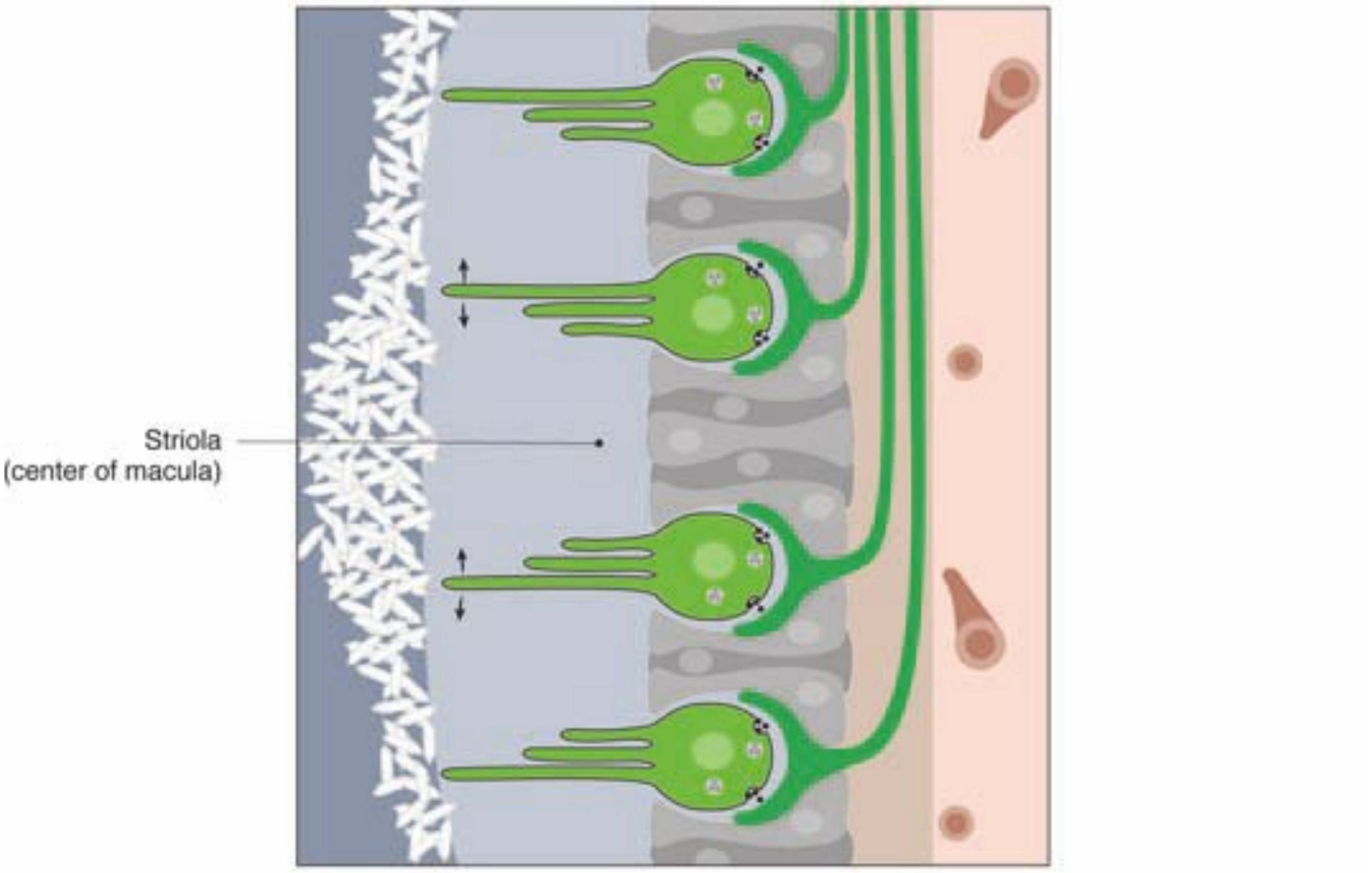


FIGURE VIII–4 Sensory transduction in hair cells.

Mechanoelectrical transduction channels (orange) are located at the tips of the shorter two rows of cilia. Tip links connect the channels to the sides of the longer cilia. At "rest", (center cell), approximately 15 percent of the channels are open. Inflow of positive ions at the tip causes membrane depolarization and release of neurotransmitter at the base of the hair cell, which in turn gives rise to action potentials in the primary sensory afferents. Movement of the stereocilia towards the tallest row (right cell) puts tension on the tip links and results in more transduction channels opening. This leads to increased depolarization of the cell membrane, increased release of neurotransmitter at the base and an increase in action potential traffic. Movement of the stereocilia away from the tallest row (left cell) closes the transduction channels, hyperpolarizes the cell and results in a decrease in neurotransmitter release and in action potential traffic. (The size of the stereocilia and their degree of movement during transduction has been exaggerated for illustrative purposes.)



UTRICLE



SACCULE

FIGURE VIII–5 Macular hair cells of the utricle and saccule

the cell, increasing membrane depolarization, which results in the release of more neurotransmitter and an increase in the impulse traffic along the sensory neurons. When the cilia sway in the opposite direction, the ion channels close, decreasing influx of potassium and hyperpolarizing the hair cells. This leads to a decrease in neurotransmitter release and a drop in the frequency of action potentials transmitted to the brain. The changing action potential traffic keeps the brain constantly informed of ongoing activities of the hair cells.

VESTIBULAR COMPONENT

There are five sensory organs in each vestibular apparatus: one in each of the utricle and saccule and one in each of the three semicircular canals. Hair cells transduce mechanical changes in head position in all five organs.

Vestibular Hair Cells

Vestibular hair cells have a ciliary bundle of 70 to 100 stereocilia and one longer kinocilium. They are of two types: type I cells and type II cells (see Fig. VIII–3).

- Type I cells are flask-shaped with a narrow neck below the cuticular plate and a bulbous cell body. The hair cell bodies are enveloped by large cuplike sensory terminals called "calyces." The sensory axons are large and myelinated. They are thought to provide the major sensory input to the brain for balance.
- Type II cells have a more cylindrical body and are in contact with bouton-shaped sensory synapses. In addition, they receive cholinergic motor synapses from the medial vestibular nucleus in the brain stem. Their associated sensory axons are thought to play a supporting role in vestibular function, possibly by affecting end-organ sensitivity.

The Otolithic Organs: the Utricle and the Saccule

The utricle and the saccule are expansions in the membranous labyrinth. Each contains a macula (spot), where a specialized patch of endothelium is located. The endothelium includes hair cells overlain by a gelatinous sheet, called the "otolithic membrane" (from the Greek words *otos*, meaning ear, and *lithos*, meaning stone). The otolithic membrane includes millions of fine calcium carbonate particles, the otoconia, lying on its surface or embedded in its top layer (see Fig. VIII–5). The otoconia give the otolithic membrane a specific gravity that is higher that that of the endolymph.

The macula of the utricle is oriented in the horizontal plane and that of the saccule lies in the vertical plane (see Fig. VIII–5). The main functions of these sensory organs are to detect movements of the head and the position of the head relative to gravitational pull.

When the head moves, inertia of the otolithic membrane causes it to lag behind. The otolithic membrane shifts with respect to the underlying hair cells, which deflects the stereocilia and generates changes in the pattern of action potentials sent to the brain.

Gravity constantly pulls the otolithic membrane toward the ground. When the head tilts, the angles between both the utricular and the saccular maculae and the ground change, causing the otolithic membranes to shift and stimulate the underlying hair cells.

Each macula includes a striola, a specialized zone at which the orientations of the hair cells change. In the utricle, the kinocilia are orientated toward the striola. In the saccule, they are oriented away from it (see Fig. VIII–5). As a group, the hair cells of the utricle and saccule are oriented in many different directions. Movement in a given direction will, therefore, excite some hair cells, inhibit others, and have little affect on still others. As a result, every different head position and movement generates a unique pattern of signals sent to the brain via the vestibular nerve.

Specialist Comment

Because the otolithic organs respond to gravity, their function is severely compromised in conditions of changing gravitational force, for example, during attitudinal changes in an aircraft. Under these conditions, therefore, pilots are trained to ignore their own vestibular signals and to trust the aircraft instruments.

Semicircular Canals

The three semicircular canals are oriented at right angles to each other (Fig. VII–6).

Each canal has an expanded end close to the utricle called the "ampulla" (see Fig. VIII–2). The ampulla contains a transverse ridge of tissue called the "crista

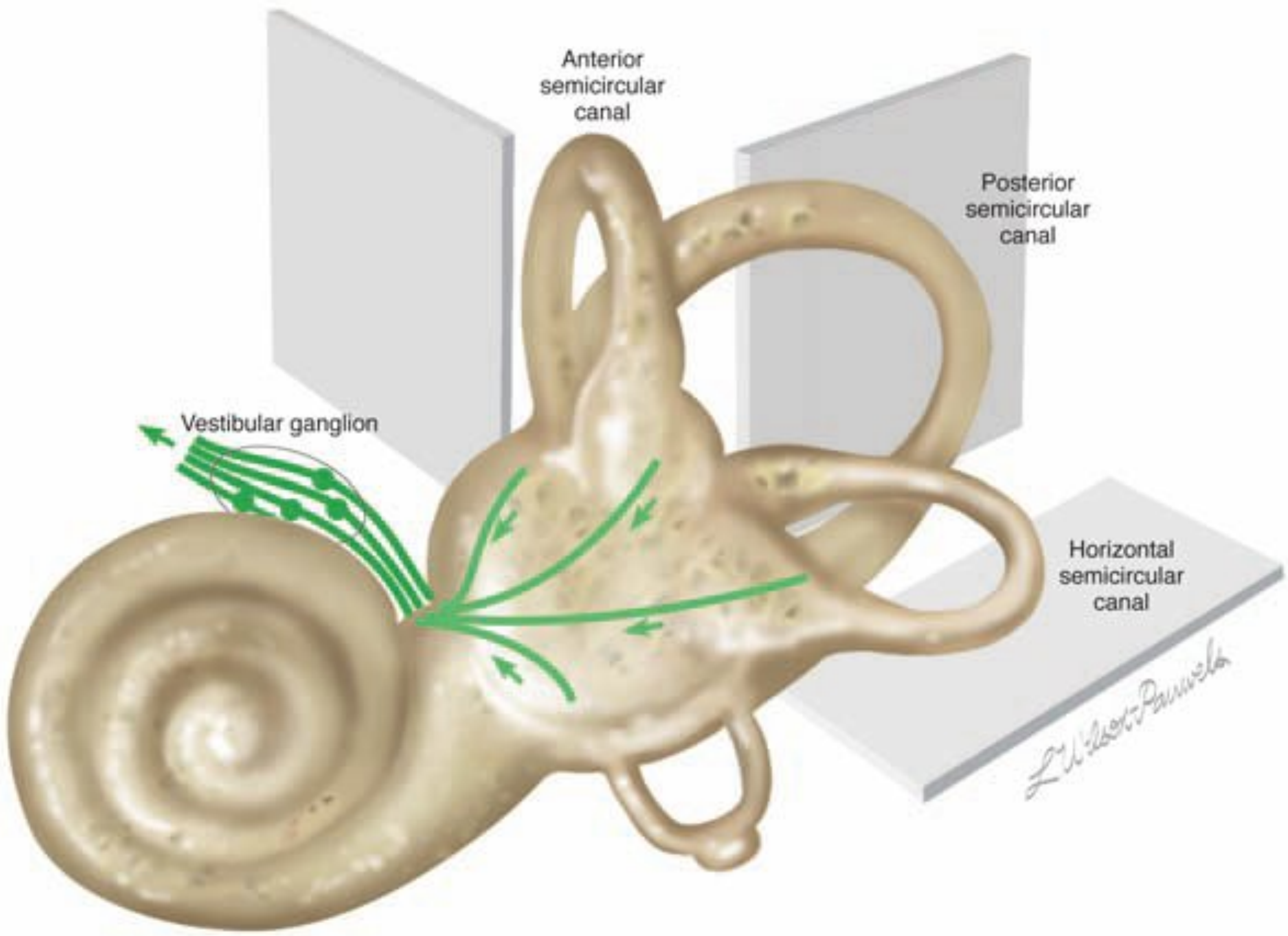

Anterior semicircular canal
Posterior semicircular canal
Vestibular ganglion
Horizontal semicircular canal
L Wilson-Pauwels

FIGURE VIII–6 Planes of the semicircular canals. Note that they are at right angles to each other.

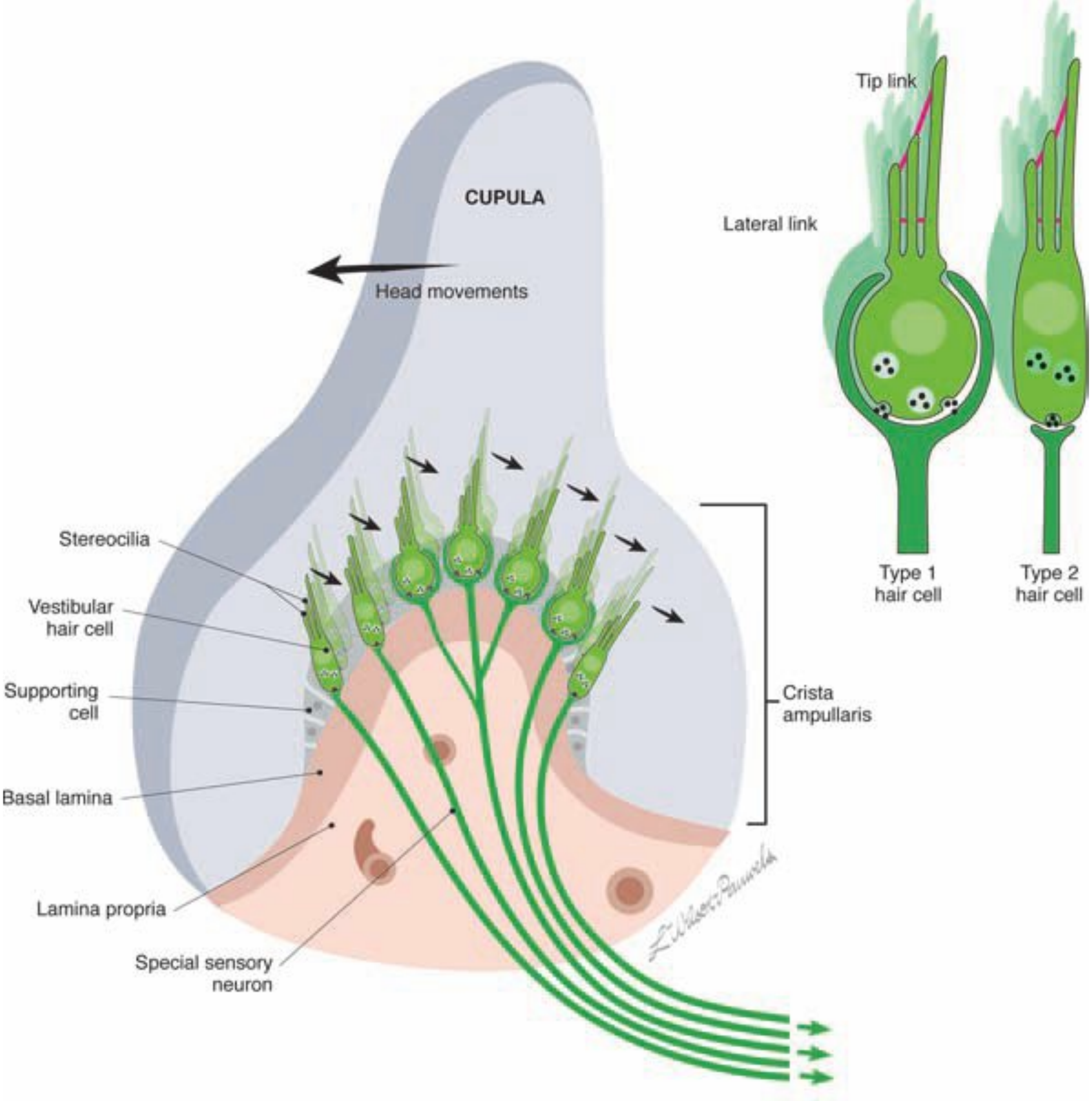


FIGURE VIII–7 Ampulla of a semicircular duct of the vestibular division of CN VIII (for clarity, structures are not drawn to scale).

ampullaris.” Hair cells are located in the epithelium of the crista and are covered by a gel-like partition called the “cupula” (Fig. VIII–7). Because the gel of the cupula does not contain otoconia, it has the same specific gravity as the endolymph and, therefore, responds only to movements of the endolymph. As the head rotates, inertia causes the endolymph within the canals to lag behind and push on the cupula. As a result, the stereocilia of the ampullary hair cells move, and the electrical properties of the hair cells change. Type I hair cells are preferentially located on the apex of the crista, where they are most affected by movement. Working together, the hair cells of all three semicircular canals on each side send signals to the brain, encoding rotatory head movement in all three planes.

Vestibular Nerve

Neurotransmitter released by the hair cells in the maculae and ampullae affects the peripheral processes of the primary sensory neurons whose cell bodies form the vestibular (Scarpa's) ganglion (see Fig. VIII–1).

The central processes of the primary vestibular neurons form the vestibular component of CN VIII. They travel with the cochlear afferents through the internal acoustic meatus to the vestibular nuclei on the dorsal surface of the brain stem at the junction of the pons and the medulla and to the floccoulo-nodular lobe of the cerebellum.

Vestibular Nuclear Complex

The vestibular nucleus (Fig. VIII–8) is composed of four major subnuclei located in the floor of the fourth ventricle at the pontomedullary junction. They are named the "superior," "medial," "lateral" (Deiter's), and "descending" (inferior) nuclei. The subnuclei mediate several functions:

The Vestibulo-ocular Reflex

When the head moves, the eyes must move at the same speed and by the same amount but in the opposite direction in order to maintain visual fixation on an object. Signals predominantly from the semicircular canals project to the superior and medial vestibular nuclei. These, in turn, send signals to the abducens, trochlear, and oculomotor nuclei via the ascending medial longitudinal fasciculus to drive the appropriate compensatory eye movements (see Fig. VIII–8) (see Chapter 13 for a description of eye movements). The otolithic organs (utricle and saccule) also have a small projection to the nuclei of oculomotor and trochlear nuclei to drive incyclotorsional and excyclotorsional movements of the eyes to compensate for head tilt. The vestibulo-ocular reflex is one of the pathways tested in the determination of brain death.

Vestibulocolic Reflex

The medial and descending nuclei form a substantial bilateral projection caudally to the cervical spinal cord via the descending medial longitudinal fasciculus (see Fig. VIII–8). Its function is to maintain a level head position while the body is moving.

Vestibulospinal Reflex

Input predominantly from the otolith organs projects to the lateral (Dieter's) nucleus, which projects ipsilaterally to the spinal cord mostly in the lateral vestibulospinal tract to coordinate postural responses to gravity. Its function is to activate the antigravity (extensor) muscles to maintain an upright stance.

All vestibular nuclei send a small number of axons via the thalamus to the somatosensory cortex, where they provide for conscious appreciation of balance and head position. Projections to the reticular formation mediate nausea and vomiting, as anyone

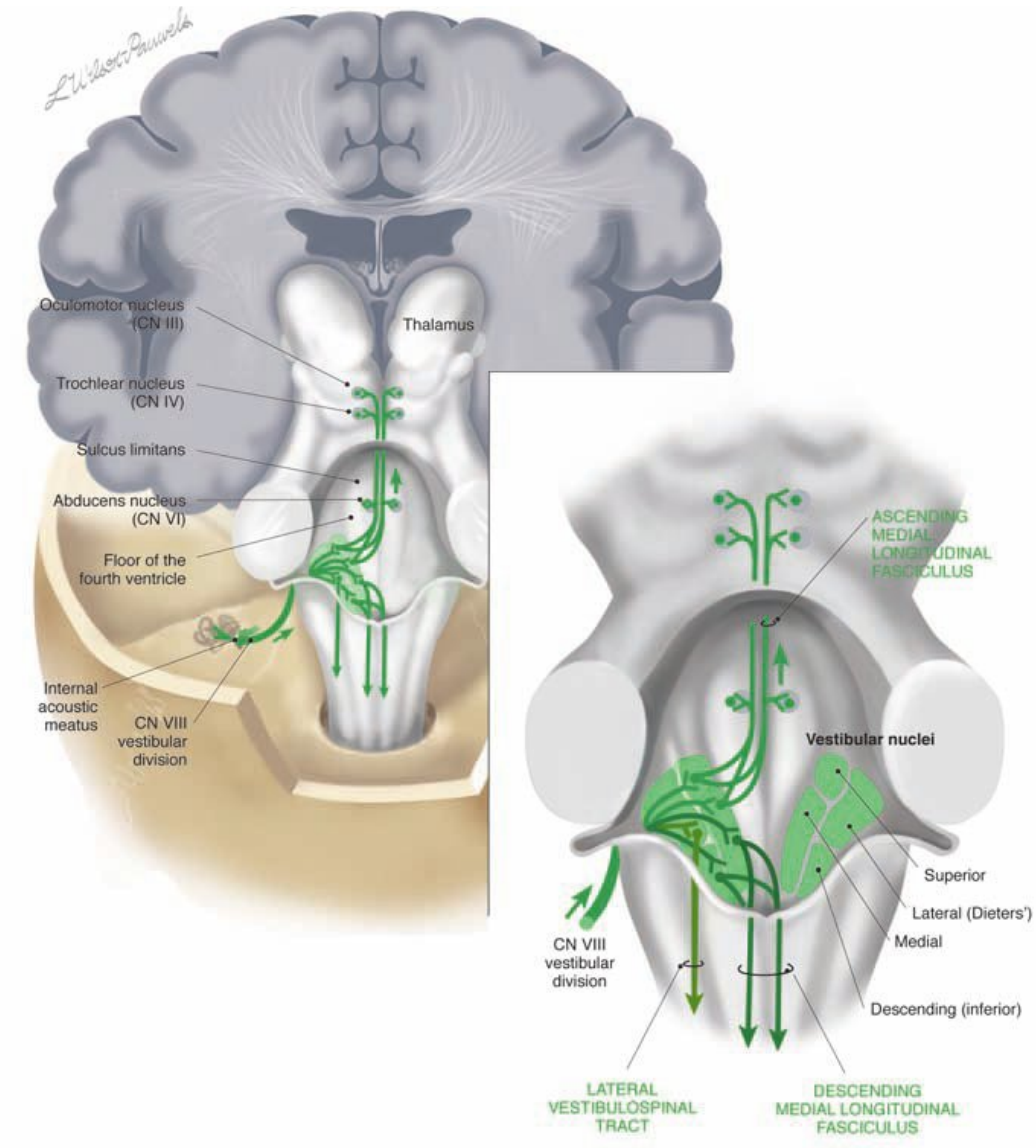

L Wilson-Pauwels
Oculomotor nucleus (CN III)
Thalamus
Trochlear nucleus (CN IV)
Sulcus limitans
Abducens nucleus (CN VI)
Floor of the fourth ventricle
Internal acoustic meatus
CN VIII vestibular division
ASCENDING MEDIAL LONGITUDINAL FASCICULUS
Vestibular nuclei
Superior
Lateral (Dieters')
Medial
Descending (inferior)
CN VIII vestibular division
LATERAL VESTIBULOSPINAL TRACT
DESCENDING MEDIAL LONGITUDINAL FASCICULUS

FIGURE VIII–8 Central sensory pathways of the vestibular division of the vestibulocochlear nerve. Note, the size of the brain stem is exaggerated.

who has ever been seasick can confirm. Other projections to the reticular formation mediate other autonomic responses such as an increase in blood pressure to compensate for the drop in blood pressure when one changes position from lying or sitting to standing. Figure VIII–8 illustrates only the major projections from the nucleus.

COCHLEAR COMPONENT

The ability to hear is extremely important for survival. Sound has two basic characteristics that humans must be able to discriminate: intensity (volume) and frequency (pitch). Intensity and frequency are encoded in the cochlea by means of its mechanical and electrical characteristics. Detecting the location of a sound source is also important for survival. Location is determined by the central nervous system. Sound waves in the air enter the external acoustic meatus and strike the tympanic membrane (ear drum), causing it to vibrate. The vibrations are carried through the middle ear cavity by a chain of three small bones, the malleus, the incus, and the stapes, to the oval window of the cochlea (Fig. VIII–9). The middle ear mechanism amplifies the force applied to the oval window.

Anatomy of the Cochlea

The cochlea is the spiral part of the bony labyrinth that houses the cochlear duct. The axis of the spiral sits in the horizontal plane of the body at an angle of approximately 45 degrees to the sagittal plane. The tunnel spirals two and a half turns around a bony center called the “modiolus” (see Figs. VIII–1 and VIII–2). The modiolus is honeycombed with spaces that accommodate the spiral ganglion cells and fibers of the cochlear nerve. The cochlea communicates with the middle ear cavity via two openings in the bone: the oval window (fenestra vestibuli), covered by the foot plate of the stapes, and the round window (fenestra cochlea), covered by a thin, flexible diaphragm (the secondary tympanic membrane; see Fig. VIII–9).

The cochlear duct, part of the membranous labyrinth, is attached to the walls of the cochlea such that it divides the cochlear space into three compartments (see Figs. VIII–1 and VIII–9): the scala vestibuli, the scala tympani, and the cochlear duct itself (also known as the “scala media”). The scalae vestibuli and tympani are continuous at the apex of the cochlea through an opening called the “helicotrema.” The cochlear duct contains the organ of Corti (see Fig. VIII–9). The organ of Corti is a long (>3 cm) ribbon-like structure that sits on the basilar membrane.

Cochlear Hair Cells

Cochlear hair cells are also of two types: inner hair cells and outer hair cells (Fig. VIII–10). In the human, they include 30 to 300 cilia arranged in three or four rows from shortest to longest on the cuticular plate. They do not have a kinocilium, at least in the adult.

- Inner hair cells are large and are arranged in a single row in the cochlea. The cilia of the inner hair cells are arranged in relatively straight rows across the cuticular plate, with the shortest of the cilia facing the center of the cochlea. Each inner hair cell sends signals to the brain via 15 to 20 dedicated sensory neurons. The inner hair cells synapse with more than 90% of the cochlear sensory neurons and, therefore, are primarily responsible for hearing.

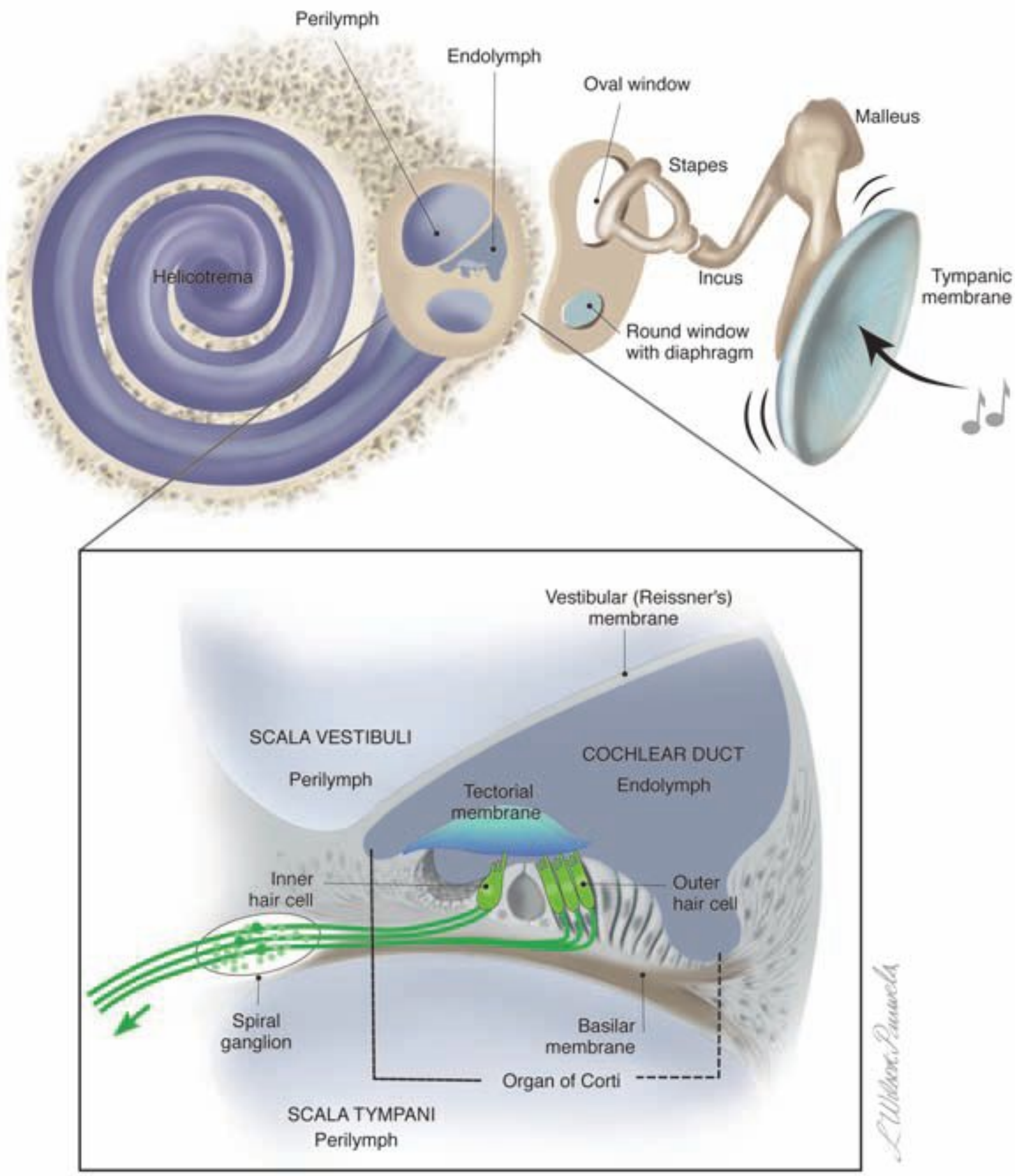


FIGURE VIII–9 Membranous labyrinth of the cochlear duct. The organ of Corti sits on the basilar membrane within the cochlear duct.

- Outer hair cells are smaller and three to four times more numerous than inner hair cells. They are arranged in three parallel rows in the cochlea, peripheral to the inner hair cells (i.e., further from the center of the cochlea). Their cilia are arranged in rows that have a characteristic “U” or “W” shape across the cuticular plate, with the shortest cilia facing the center of the cochlea. They provide less than 10% of the auditory sensory traffic to the brain; however,

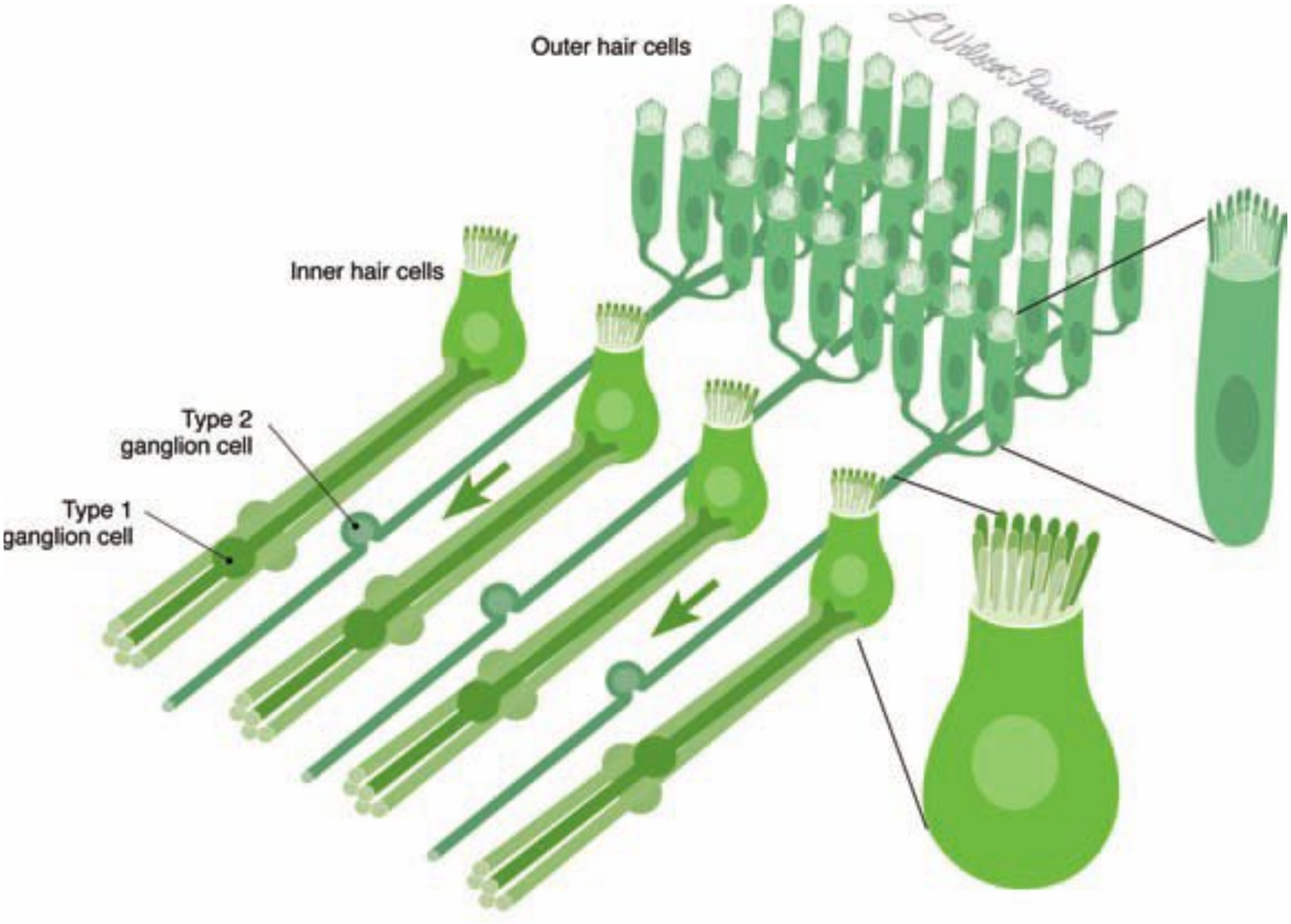


FIGURE VIII–10 Special sensory output from the hair cells of the organ of corti.

they have the unique ability to change shape with changing membrane voltage. They play a critical role in hearing by amplifying the signal to the inner hair cells and by sharpening pitch discrimination in the cochlea (see "The Cochlear Amplifier," later).

The inner and outer hair cells are arranged in rows along the length of the organ of Corti. The inner hair cells (3500) form a single row close to the modiolus and the outer hair cells (12,000–14,000) form three rows farther away from the modiolus. The tectorial membrane, a ribbon-like strip of gel, extends from the modiolar attachment of the cochlear duct into the center of the duct, where it overlays the hair cells. The tallest of the stereocilia of the outer hair cells are embedded in the base of the tectorial membrane (see Fig. VIII–9).

Function of the Cochlea

Movements of the stapes in the oval window set up pressure waves within the perilymph, which cause the round window diaphragm to move in the opposite direction. If the round window and its diaphragm were not present, movement of the stapes would be prevented by the incompressible nature of the fluid in the labyrinth. The round window, with its flexible diaphragm, allows the fluid to move slightly, permitting propagation of pressure waves along the basilar membrane.

Inner hair cells are situated along the length of the basilar membrane. When part of the basilar membrane is displaced by sound, local currents are set up in the overlying endolymph. The fluid movement activates the inner hair cells in the same location. The activated inner hair cells stimulate their associated primary sensory neurons and sound signals are sent to the brain. Because the frequency of the sound determines the location of basilar membrane movement, it also determines which population of inner hair cells respond. A given population of inner hair cells, therefore, will always signal the same frequency.

The intensity (volume) of the sound is signaled in two ways: (1) by the number of sensory neurons firing and (2) by the frequency of the action potentials transmitted to the brain. The more axons that are firing and the higher the number of action potentials, the more intense is the perceived sound.

Most sound in our environment is complex, that is, it consists of several tones at different frequencies and intensities. Under normal circumstances, several areas of the basilar membrane, and therefore, several groups of inner hair cells, are activated at once.

The Cochlear Amplifier

The transduction mechanism described previously takes place in an aqueous environment. The dampening effect of the cochlear fluids and mechanical resistance inhibits the movement of the basilar membrane. The attenuation is significant enough that the cochlea would be unresponsive at lower sound intensities if not for the amplifying activity of the outer hair cells.

Outer hair cells respond to sound in the same way that inner hair cells do, in that movement of the stereocilia causes changes in the electrical charge across the cell membrane resulting in changes in neurotransmitter release. However, outer hair cells have an additional response that inner hair cells do not. Outer hair cells have the unique ability to become shorter or taller in response to changes in membrane potential.

The longest stereocilia of the outer hair cells are anchored in the tectorial membrane. When the basilar membrane moves upward, the outer hair cells are also elevated, which creates a shearing force on their stereocilia. As a result, the cilia bend outward and the cells depolarize. Depolarization of the membrane causes the outer hair cells to become shorter, and they pull the basilar membrane up even higher. When the basilar membrane moves downward, the opposite happens. The outer hair cells hyperpolarize, elongate, and push the basilar membrane down even farther. The outer hair cells, therefore, amplify the movements of the basilar membrane in the same way that pushing someone on a swing will make the swing go higher. In turn, the amplified movements of the basilar membrane increase the stimulation of the adjacent inner hair cells, enhancing their response to sound.

Central Pathways

Sound Perception Pathway

In the auditory system, the sensory pathway from the periphery to the cerebral cortex is much more complex than in other sensory pathways. Primary sensory axons from the spiral ganglion project to the cochlear nucleus in the brain stem. The cochlear nucleus is located at the junction of the pons and medulla and is draped over the inferior cerebellar peduncle (Fig. VIII–11). Functionally, it is divided into the dorsal and ventral cochlear nuclei.

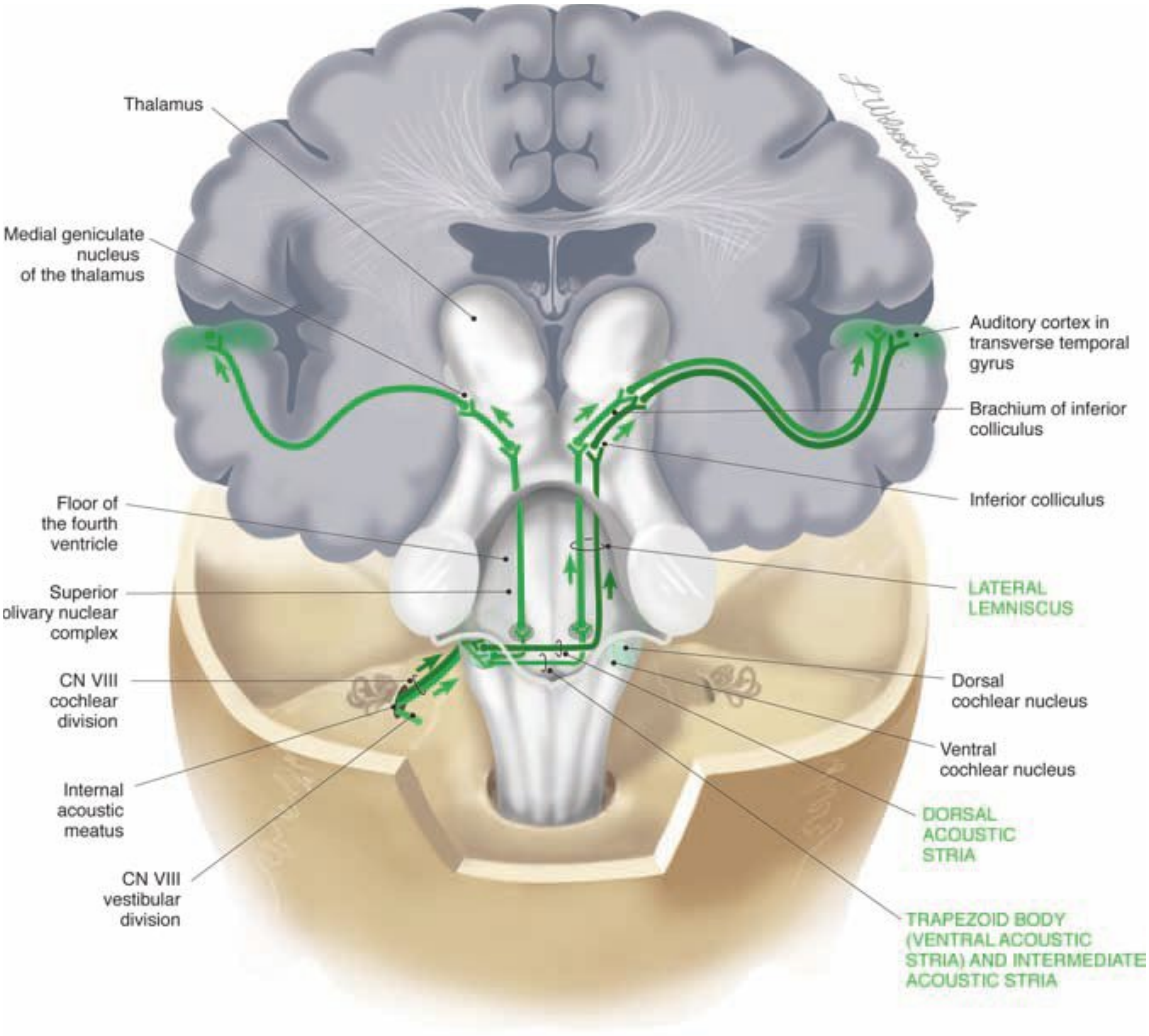

Thalamus
Medial geniculate nucleus of the thalamus
Auditory cortex in transverse temporal gyrus
Brachium of inferior colliculus
Inferior colliculus
Floor of the fourth ventricle
LATERAL LEMNISCUS
Superior olivary nuclear complex
Dorsal cochlear nucleus
CN VIII cochlear division
Ventral cochlear nucleus
Internal acoustic meatus
DORSAL ACOUSTIC STRIA
CN VIII vestibular division
TRAPEZOID BODY (VENTRAL ACOUSTIC STRIA) AND INTERMEDIATE ACOUSTIC STRIA

FIGURE VIII–11 Central afferent pathways of the cochlear division of the vestibulocochlear nerve. Note that the size of the brain stem is exaggerated. (Illustration is based on Barr 2009, Figure 21.6.)

Neurons in the dorsal cochlear nucleus project across the midline via the dorsal acoustic stria and then rostrally in the lateral lemniscus to synapse in the contralateral inferior colliculus (see Fig. VIII–11). Neurons in the ventral cochlear nucleus project bilaterally to the superior olivary nuclear complex. Superior olivary nuclear complex neurons project via the lateral lemnisci to the inferior colliculi.

The inferior colliculus projects via the brachium of the inferior colliculus to the medial geniculate nucleus of the thalamus and from there to the transverse temporal gyri on the superior surface of the temporal lobe where sound is perceived. Because the pathway from the cochlear nuclei to the auditory cortex is bilateral, lesions in the brain stem do not produce hearing deficits confined to one ear. The tonotopic arrangement of the axons in the hearing pathways extends to the primary auditory cortex. High-pitch sounds are perceived in the medial aspects of the transverse temporal gyrus, and low-pitch sounds are perceived in the lateral aspects.

Auditory Reflexes

Sound Localization. Sound coming from one side of the head will arrive sooner at, and its intensity will be slightly higher in, the ipsilateral ear. Cells in the superior olivary nuclei receive signals from both ears and, therefore, play a major role in the localization of sound. They compare the time of arrival and the intensity and send this information to higher centers for sound localization.

Stapedial Reflex. Intense sound damages the hair cells in the cochlea and they do not regenerate. The intensity of sound entering the cochlea is decreased by an order of magnitude by the contraction of the stapedius muscle (CN VII) in the stapedial reflex. Sound from either or both ears projects to both superior olivary nuclei, from which signals are sent to both facial nuclei to drive contraction of the stapedius muscles in both ears. Unfortunately, the onset of muscle contraction does not occur quickly enough to protect the cochlea from the damaging effects of percussive sounds such as hammering or gunshots. Furthermore, the reflex depends on muscle action and, therefore, will fatigue after prolonged use.

Motor Olivocochlear Pathway. The inhibitory pathway to the outer hair cells, which inhibits electromotility, arises in the superior olivary complex. The role of this pathway is thought to be in sharpening pitch perception.

CASE HISTORY GUIDING QUESTIONS

1. Why did Paul experience “ringing” in his ears before he saw his doctor?
2. What are other causes of hearing loss?
3. In CN VIII lesions, can there be impairment of vestibular function without hearing loss or vice versa?
4. What other CNs can be involved when there is a tumor of CN VIII?

1. Why did Paul experience “ringing” in his ears before he saw his doctor?

The perception of ringing in the ears in the absence of an external sound source is called “tinnitus” (from the Latin *tinnire*, to ring). Ten to fifteen percent of the population experience tinnitus, and for many, it can be bothersome enough that they seek medical help. It is generally accepted that the ringing sensation is due to hyperexcitability in the central hearing pathway, most commonly in the primary auditory cortex and inferior colliculus, in response to sensorineural hearing loss in the cochlea or auditory nerve. In Paul’s case, his auditory nerve damage caused the increased activity in his central auditory pathway. Tinnitus is more common in the elderly, who have presbycusis (hearing loss related to aging). Temporary tinnitus can be caused by high doses of acetyl salicylic acid, which inhibits the cochlear amplifier.

2. What are other causes of hearing loss?

Hearing loss can occur as a result of a lesion or disease process anywhere along the course of the auditory pathway from the auditory apparatus to the auditory cortex. The tympanic membrane, ossicles, and cochlea can be damaged by trauma or infection. Interference with the transmission of sound to the cochlea is defined as a **conductive** hearing loss. The organ of Corti and/or the auditory nerve may be damaged by noise exposure, infections, toxic drug exposure, or tumors. The central auditory pathways can be affected by strokes, multiple sclerosis, or tumors. Interference with the transduction mechanism or the transmission of impulses to the auditory cortex is defined as a **sensory** hearing loss. Damage to the transmission mechanism, transduction mechanism, or the auditory nerve results in hearing loss only on the affected side. Within the central nervous system, however, hearing signals are carried bilaterally in the lateral lemniscus and represented bilaterally in the auditory cortex. Unilateral lesions in the central nervous system, therefore, do not usually result in hearing loss in the ear on the affected side. In fact, total removal of one cerebral hemisphere of the brain in humans does not result in any major change of auditory sensitivity in either ear.

3. In CN VIII lesions, can there be impairment of vestibular function without hearing loss or vice versa?

Although referred to as the “vestibulocochlear nerve,” the nerve is actually two distinct nerves, vestibular and cochlear, that travel together. It is, therefore, possible for only one of the components to be involved in a disease process, but because of the close proximity in their peripheral course through the internal auditory meatus and across the cerebellopontine angle, both are usually involved simultaneously.

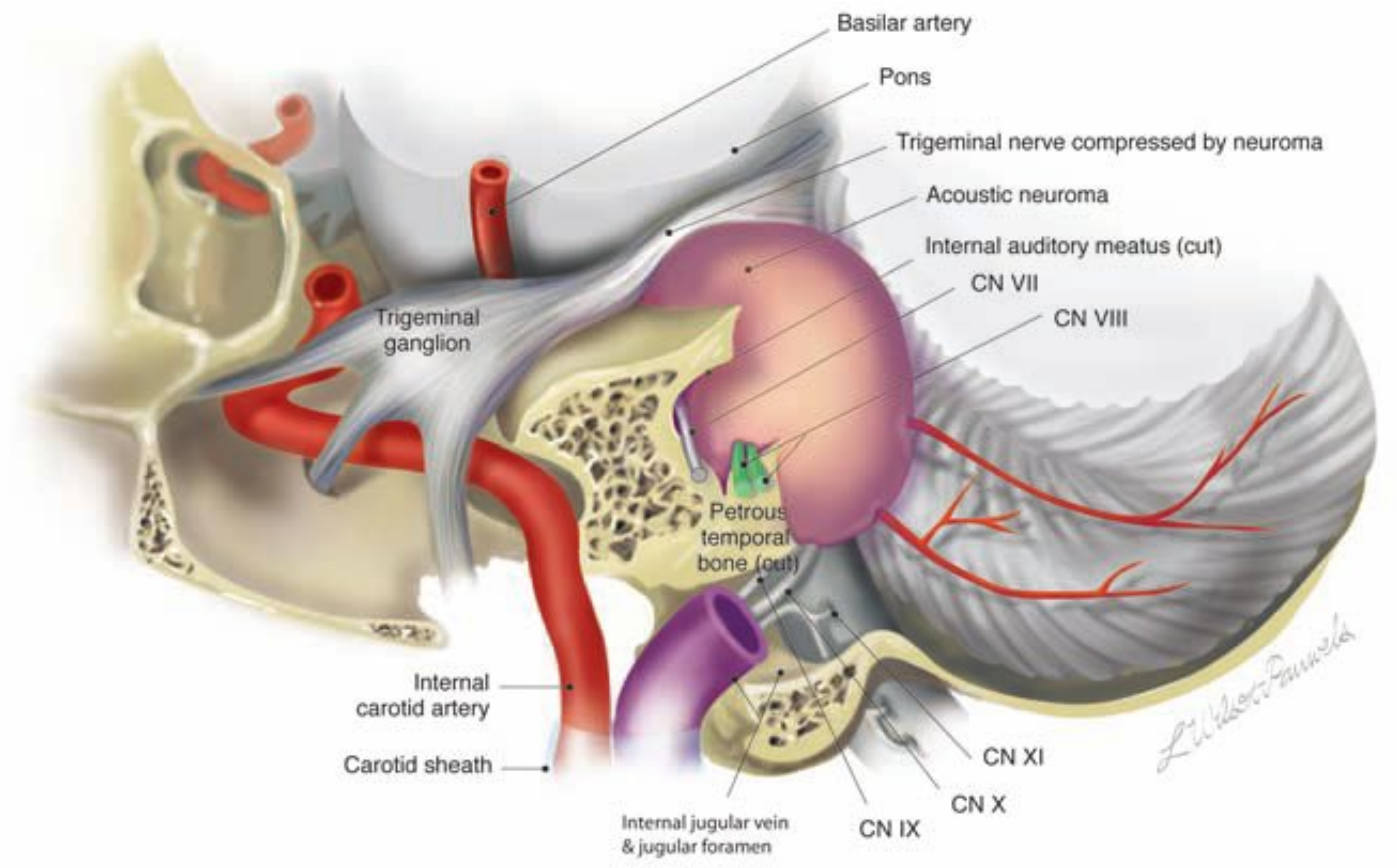

Basilar artery
Pons
Trigeminal nerve compressed by neuroma
Acoustic neuroma
Internal auditory meatus (cut)
CN VII
CN VIII
Trigeminal ganglion
Petrous temporal bone (cut)
Internal carotid artery
Carotid sheath
Internal jugular vein & jugular foramen
CN IX
CN X
CN XI
L Wilson-Pauwels

FIGURE VIII–12 An enlarged tumor (schwannoma) in the cerebellopontine angle compromising cranial nerves V, VII, and the vestibular and cochlear divisions of VIII (illustrated in green). This is a sagittal section through the jugular foramen.

Once the fibers enter the brain stem and synapse with their nuclei, the axons take different courses and are less likely to be affected simultaneously.

4. What other CNs can be involved when there is a tumor of CN VIII?

Tumors such as schwannomas and meningiomas typically compress CN VIII at the cerebellopontine angle. Other CN involvement is a reflection of the close proximity of these nerves to CN VIII. CN VII is most often affected because it traverses the internal auditory meatus and the cerebellopontine angle beside CN VIII (see Fig. VIII–12). Facial nerve damage results in ipsilateral facial paralysis. CN V can also be involved resulting in facial numbness, tingling, and sometimes facial pain. As the tumor enlarges, it can also compress CNs IX and X.

In addition to the peripheral CNs, an expanding cerebellopontine angle tumor may compress the brain stem and interfere with transmission of sensory and motor signals between the cerebrum and the spinal cord. The magnetic resonance imaging (MRI) scan (Fig. VIII–13) shows significant compression of his brain stem and cerebellum.

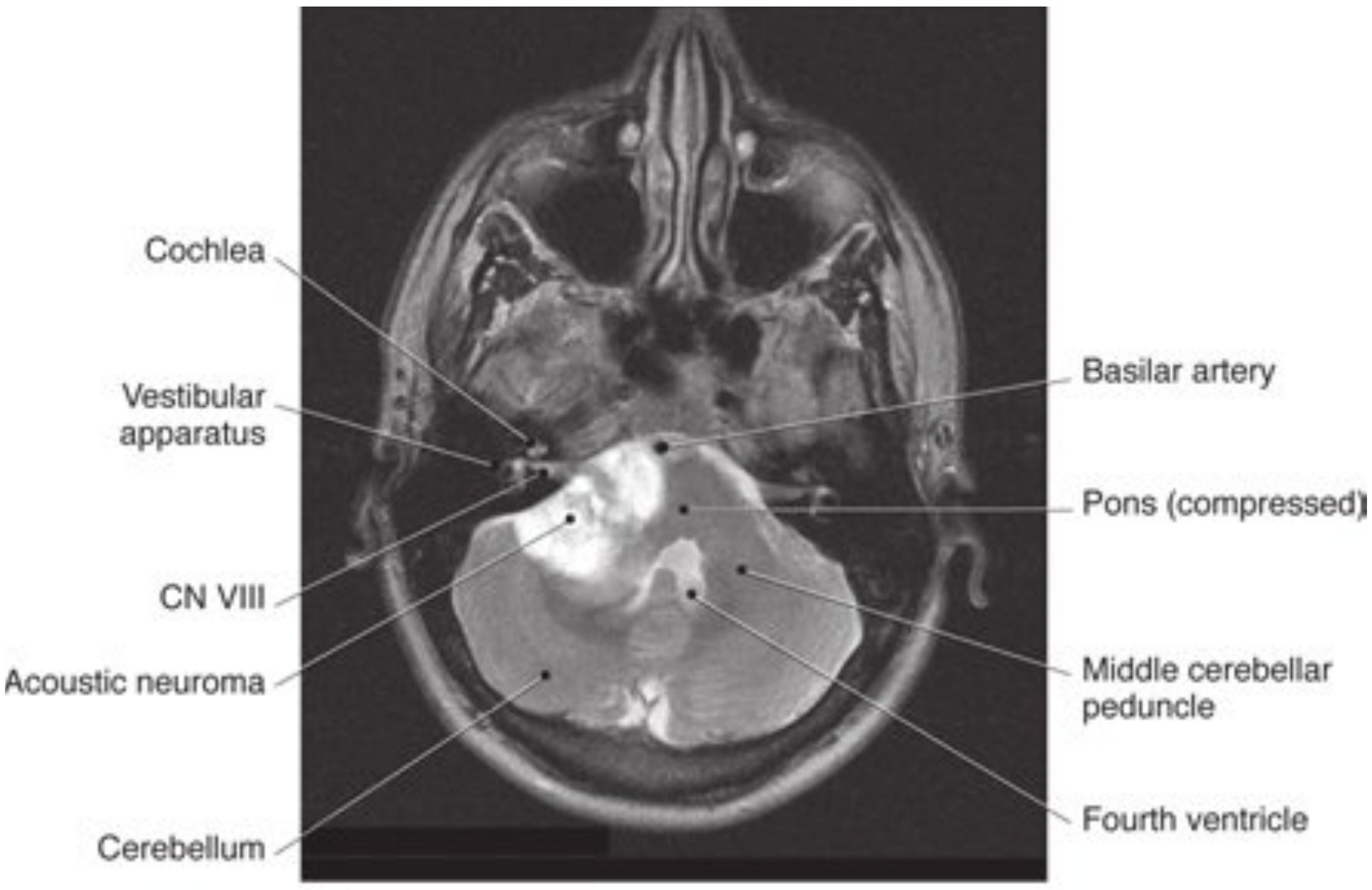


FIGURE VIII–13 Magnetic resonance image showing a large acoustic neuroma on his right vestibulocochlear nerve (left side of the image). (Courtesy of Dr. D. S. Butcher, William Osler Health Centre).

CLINICAL TESTING

Observation of the Vestibular Nerve

Because the vestibular nerve affects both eye and postural movements, the function of the nerve can be evaluated by observing the patient's eye movements and postural balance. As the emergency physician noted, Paul had horizontal nystagmus. Jerk nystagmus is the most common form and involves movements that alternate between a slow smooth phase and a fast (jerk) corrective phase. (See also "Cranial Nerves Examination" on website.)

Caloric Testing of the Vestibular Nerve

Integrity of the vestibular nerve and its end organs can also be assessed using caloric stimulation testing. This is not performed as a routine part of the bedside cranial nerve testing but is done if vestibular nerve impairment is suspected. Caloric testing involves irrigation of the external auditory canal with warm and cold water. The patient is tested with the head of the bed at 30 degrees from the horizontal. This position brings the horizontal semicircular canals into a more vertical plane, which is a position of maximal sensitivity to thermal stimuli. Using cool water (relative to body temperature) at 30°C or lower, the external auditory canal is irrigated for 30 seconds. In a normal individual, the eyes tonically deviate to the irrigated side followed by nystagmus to the opposite side. The nystagmus appears after a latent period of about 20 seconds and persists for 1.5 to 2 minutes. The same procedure is repeated after approximately 5 minutes using warm water at 44°C or higher. When warm water is used, nystagmus is toward the irrigated ear. These results can be summarized using the mnemonic COWS, Cold Opposite Warm Same. (See also "Cranial Nerves Examination" on website.)

Simple Hearing Test for the Cochlear Nerve

Auditory stimuli (hearing) can be assessed easily at the bedside. The patient covers one ear and the examiner whispers into the patient's other ear and asks the patient to repeat what has been whispered. The examiner can increase the volume if the patient fails to hear. This is repeated on the other side and the two sides are compared.

If reduced hearing is detected, the next step is to determine whether this is due to a conductive or a sensorineural loss. Conductive loss implies an obstruction in the transmission of sound from the air to the cochlea. This could be due to wax, an ear infection, or could be caused by damage to the tympanic membrane or the ossicles. A sensorineural loss, conversely, implies damage to the auditory pathway anywhere from the cochlea to the auditory cortex. The two types of hearing loss can be distinguished by the use of the Rinne's and Weber's tests. (See also "Cranial Nerves Examination" on website.)

Rinne's Test for the Cochlear Nerve

The Rinne test involves placement of a vibrating 512-Hz tuning fork on the mastoid process (Fig. VIII–14). Sound from the tuning fork is carried to the cochlea via bone conduction, bypassing the middle ear amplification mechanism. When the patient can no longer hear the ringing sound, the fork is removed from the mastoid process and brought to within 2.5 cm of the external auditory meatus to permit air to conduct sound through the external and middle ear to the cochlea. A patient with normal hearing will be able to hear the tuning fork again. This constitutes a positive Rinne test. If an individual fails to hear the ringing when the fork is placed alongside the ear, this is a negative test and implies that there is a conductive defect with the problem located in the external or middle ear.

Weber's Test

Weber's test can help distinguish between conductive deafness and sensorineural deafness. A vibrating 512-Hz tuning fork is placed in the middle of the forehead (Fig. VIII–15). Sound is transmitted to the cochlea by bone conduction, bypassing the middle ear amplification system. In a person with normal hearing, the sound is heard equally in both ears. If an individual has sensorineural hearing loss, the ringing will not be as loud on the affected side. If an individual has a conductive hearing loss, the ringing will be louder on the affected side.

It is not immediately obvious why Weber's test would result in the perception of louder sound in an ear with conductive hearing loss. The explanation is that in a normal ear, sound from the tuning fork arrives at the cochlea via two pathways:

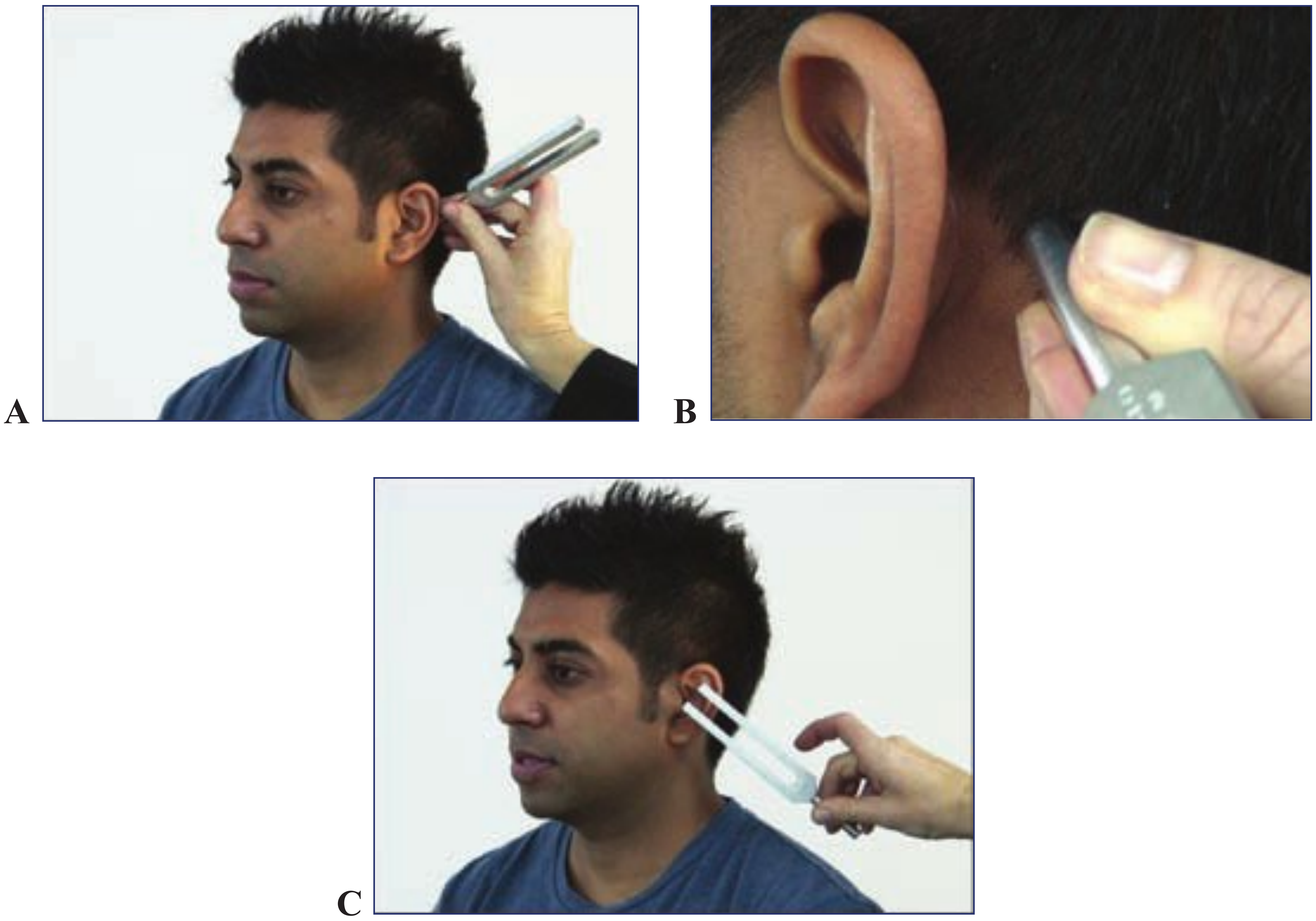


FIGURE VIII–14 The Rinne test to evaluate hearing.
A. Vibrating tuning fork (512 Hz) is placed on the mastoid process.
B. Enlargement of A.
C. Tuning fork is brought to within 2.5 cm of the external auditory meatus.

through the bony structures of the skull and through the air via the external ear. Because of the differences in pathway length and speed of sound conduction through different media, the sound waves via each pathway arrive at the cochlea somewhat out of phase. The out-of-phase sound waves attenuate each other and the sound is perceived as less intense. When the conductive pathway is blocked, the attenuation does not occur and sound arriving through bone conduction is at full strength. This

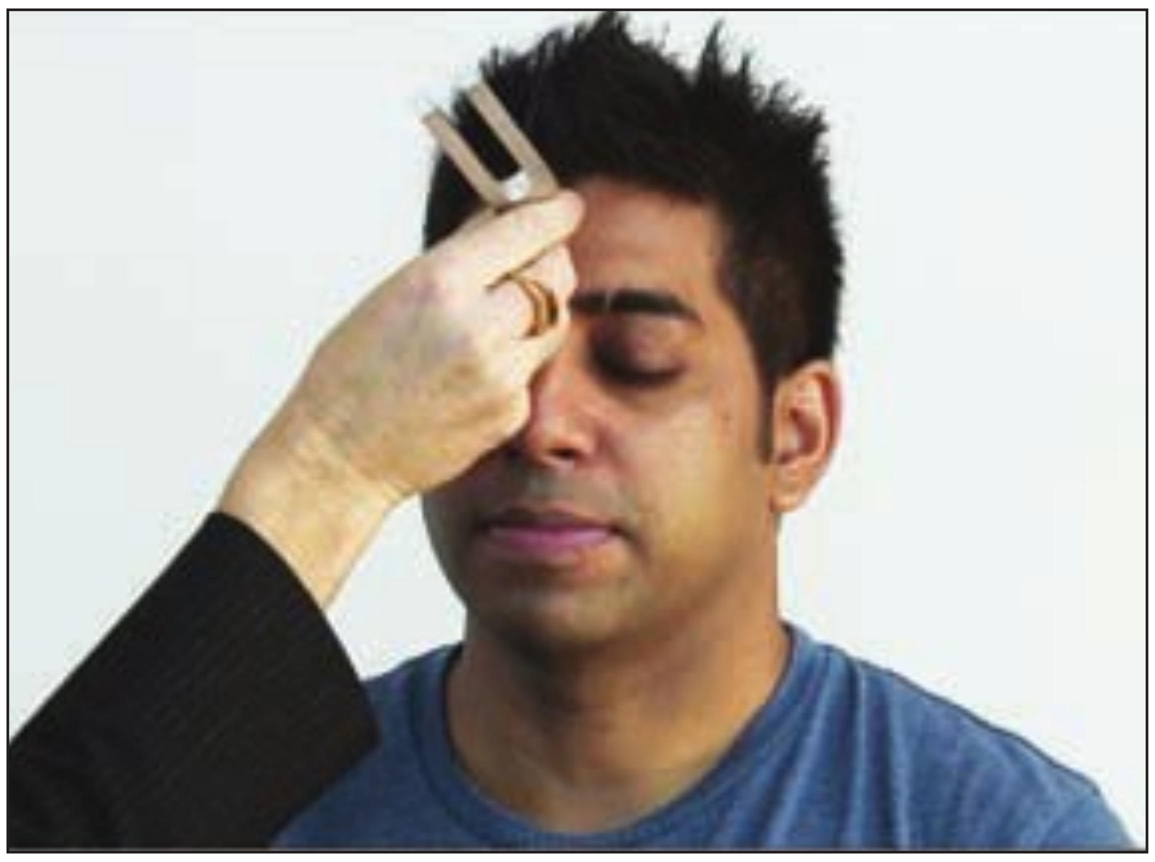


FIGURE VIII–15 Weber's test to distinguish between conductive and sensorineural deafness. A vibrating tuning fork (512 Hz) is placed in the middle of the forehead.

effect is particularly noticeable at low (≤512-Hz) frequencies; therefore, Weber's test can be performed effectively only using a low-frequency tuning fork.

You can simulate a Weber's test on yourself. Hum a note. The sound from your larynx will be carried to your cochlea by bone and air conduction. While humming, alternately block and unblock one external auditory meatus, creating a temporary conduction block. Note the increase in perceived loudness in the blocked ear.

ADDITIONAL RESOURCES

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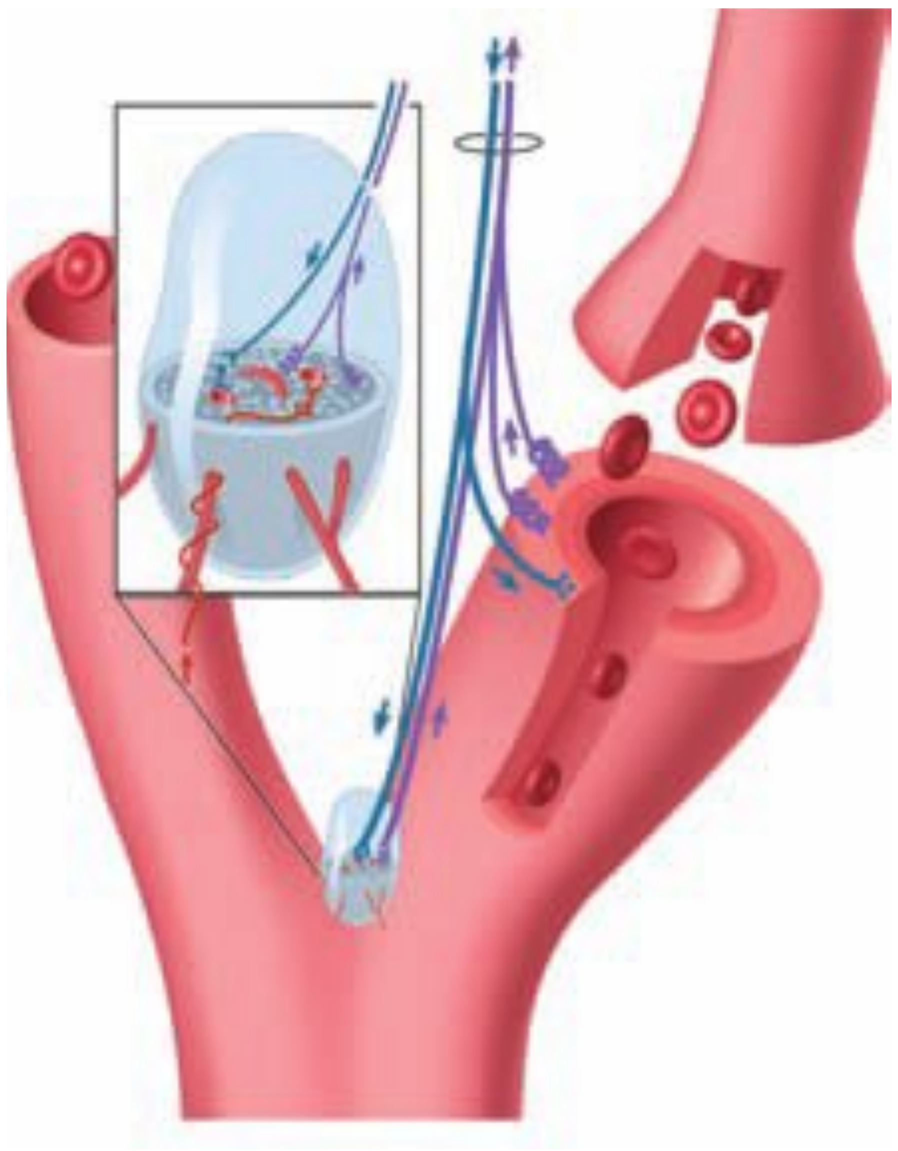

IX

Glossopharyngeal Nerve

CASE HISTORY

Allen is a 43-year-old construction worker. While he was eating dinner, he developed pain in the left side of his throat. The pain was short, brief, and stabbing in nature and only occurred when he swallowed. Allen was eating fish at the time and assumed that a fish bone was lodged in his throat. He went to the emergency department and was assessed by an otolaryngologist, but no abnormalities were found. It was suspected that a fish bone had scratched Allen's throat and that his symptoms would soon resolve.

A month later, Allen's pain was still present; in fact, it had increased. The pain occurred not only when he swallowed but also when he spoke and coughed. Again, while eating dinner, Allen experienced the same pain with swallowing. He got up from the table, but after a few steps, he collapsed unconscious to the floor.

Although Allen quickly regained consciousness, he was immediately taken to the emergency department where he was put on a heart rate and blood pressure monitor to evaluate the function of his cardiovascular system. The emergency room physician noticed an interesting correlation. Whenever Allen swallowed, he had a paroxysm (a sudden recurrence or intensification of symptoms) of pain followed within 3 to 4 seconds by a decrease in heart rate (bradycardia) and a fall in blood pressure (hypotension). Allen was examined by a neurologist who noticed no evidence of reduced perception of pharyngeal pinprick touch, or pharyngeal motility. A diagnosis of glossopharyngeal neuralgia was made. Allen was treated with carbamazepine and his attacks of pain stopped and his cardiovascular function returned to normal.

ANATOMY OF THE GLOSSOPHARYNGEAL NERVE

The name of the glossopharyngeal nerve indicates its distribution (i.e., to the glossus [tongue] and to the pharynx). Its primary role is to supply general sensation and taste from the posterior one-third of the tongue, soft palate, and pharynx and visceral sensation from the carotid body and sinus. It has a small branchial motor component to the stylopharyngeus muscle and a parasympathetic motor component to the parotid gland and carotid body and sinus. Cranial nerve (CN) IX emerges from the medulla of the brain stem as the most rostral of a series of rootlets that emerge between the olive and the inferior cerebellar peduncle (Fig. IX–1). The nerve leaves the cranial fossa through the jugular foramen along with CNs X and XI (Fig. IX–2). Two ganglia are situated on the nerve as it traverses the jugular foramen, the superior and inferior (petrosal) glossopharyngeal ganglia. The superior ganglion contains general sensory neurons and is often thought of as a detached part of the inferior glossopharyngeal ganglion. The inferior (petrosal) ganglion is composed of nerve cell bodies of the visceral and special sensory components of the nerve.

As the nerve passes through the jugular foramen, it gives rise to six terminal branches. These are the tympanic, the carotid, the pharyngeal, the tonsilar, the lingual, and the muscular branches (see Fig. IX–1).

TABLE IX–1 Components, Nuclei, Ganglia/cells, and Functions of the Glossopharyngeal Nerve (CN IX)

Component	Nucleus	Ganglion & Cell of Origin	Function
General sensory (afferent)	Spinal trigeminal nucleus	Superior glossopharyngeal ganglion	For general sensation from the posterior one-third of the tongue, tonsil, soft palate, fauces, uvula, mucosa of the internal surface of the tympanic membrane and cavity, mastoid air cells, auditory tube, and upper pharynx
Visceral sensory (afferent)	Nucleus of the tractus solitarius—middle part	Inferior glossopharyngeal ganglion	For subconscious sensations from the carotid body (chemoreceptors) and the carotid sinus (baroreceptors)
Special sensory (afferent)	Nucleus of the tractus solitarius rostral (gustatory portion)	Inferior glossopharyngeal ganglion Taste buds	For taste sensation from the posterior one-third of the tongue
Branchial motor (efferent)	Nucleus ambiguus (rostral part)		To innervate the stylopharyngeus muscle
Parasympathetic motor (visceral efferent)	Inferior salivatory nucleus Nucleus ambiguus	Otic ganglion	To innervate the parotid gland (secretomotor and vasodilatory) To control blood vessels in the carotid body (vasodilation) and blood pressure in the carotid sinus

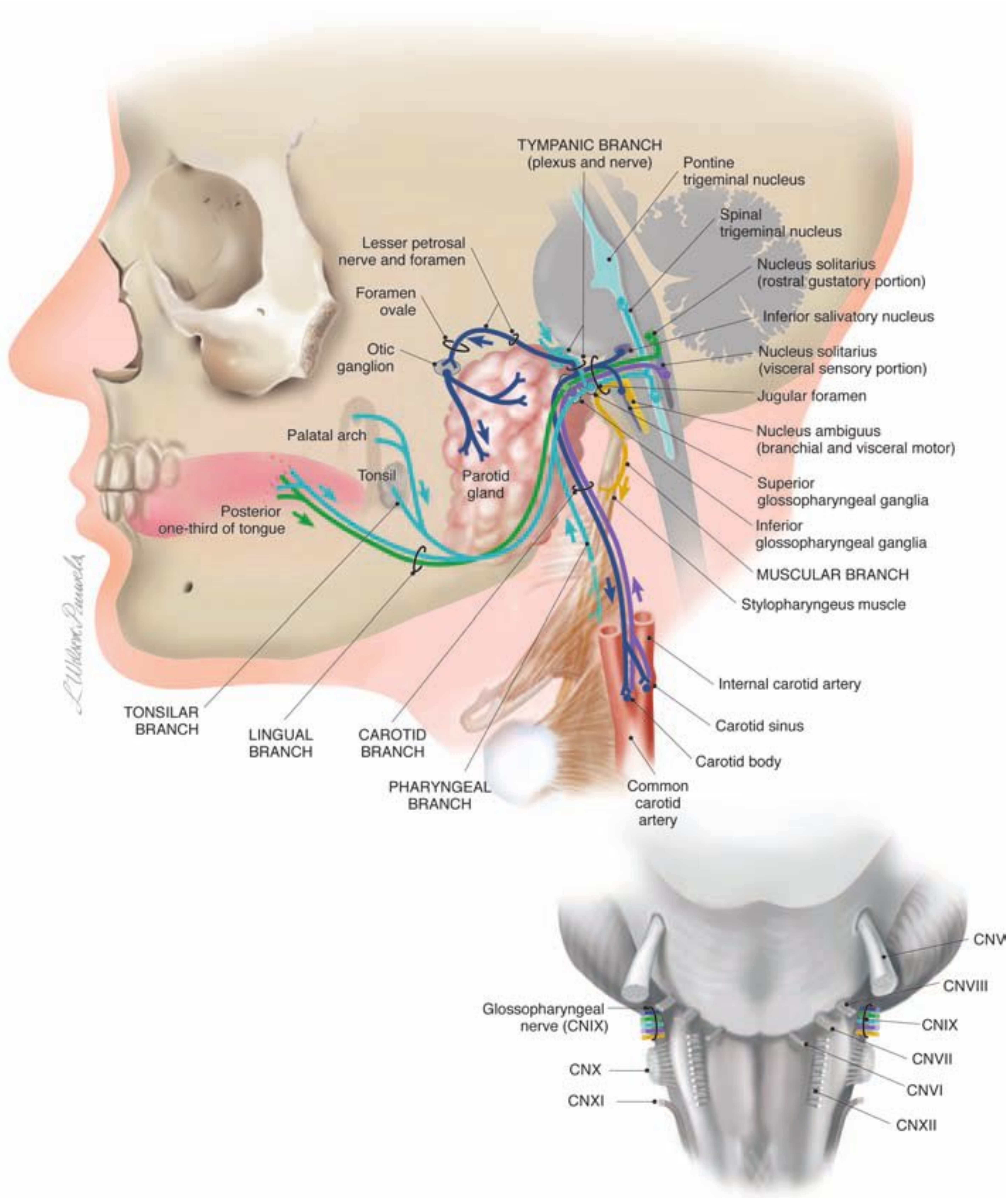

FIGURE IX–1 Overview of the glossopharyngeal nerve.

GENERAL SENSORY (AFFERENT) COMPONENT

The glossopharyngeal nerve carries general sensory signals from the posterior one-third of the tongue, tonsils, soft palate, fauces, uvula, mucosa of the internal surface of the tympanic membranes (ear drums) and the tympanic cavities (see Fig. IX–2), mastoid air cells, auditory tubes, and upper pharynx (Fig. IX–3 and Table IX–1).

Sensory fibers carried in the pharyngeal branch converge and leave the upper pharynx by piercing the superior constrictor muscle or by passing through a space between the superior and the middle constrictor muscles. These sensory fibers join with the tonsilar and lingual branches from the tonsils, soft palate, fauces, uvula, and the posterior one-third of the tongue and continue to the superior glossopharyngeal ganglion where their nerve cell bodies are located.

The tympanic branch is formed by the union of the tympanic plexus axons and comprises general sensory and the parasympathetic fibers of the visceral motor division of the nerve (see later). The sensory fibers descend through the tiny bony canal (tympanic canaliculus) and join the main trunk of the glossopharyngeal nerve at its ganglion (see Fig. IX–2).

Axons in the tympanic nerve carrying pain signals enter the medulla and descend in the spinal trigeminal tract to end on the caudal part of the spinal trigeminal nucleus (Fig. IX–4; see also Fig. IX–3). From the nucleus, axons of secondary neurons cross the midline in the medulla and project to two distinct populations of neurons in the thalamus: neurons in the VMpo (ventral medial posterior nucleus) of the thalamus, which project to the postcentral sensory cortex (head region) for identification of the location and intensity of the pain; and neurons in the dorsal

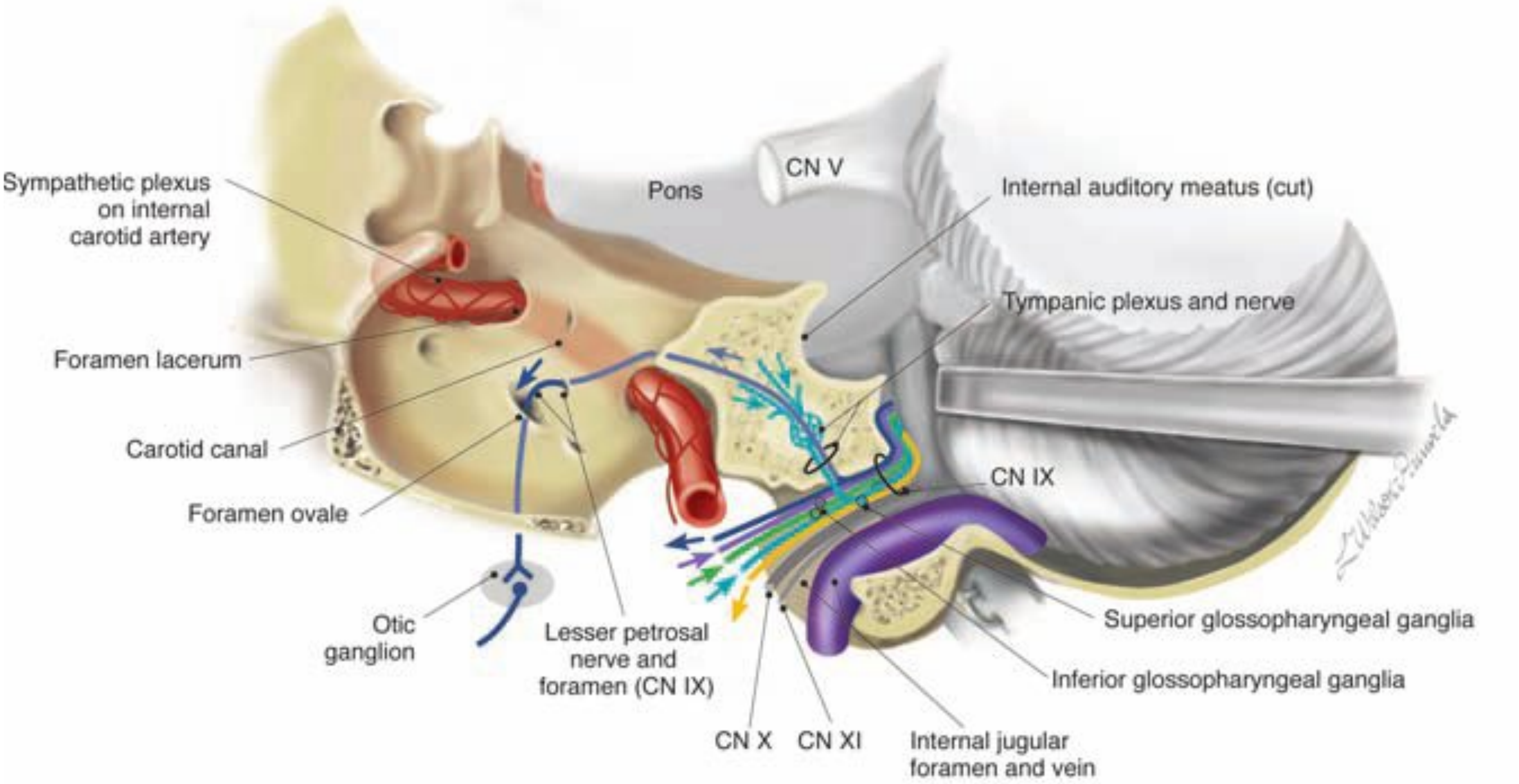
Sympathetic plexus on internal carotid artery
Pons
CN V
Internal auditory meatus (cut)
Tympanic plexus and nerve
Foramen lacerum
Carotid canal
Foramen ovale
CN IX
Otic ganglion
Lesser petrosal nerve and foramen (CN IX)
Superior glossopharyngeal ganglia
Inferior glossopharyngeal ganglia
CN X
CN XI
Internal jugular foramen and vein

FIGURE IX–2 Tympanic branch of cranial nerve IX (cut through the petrous temporal bone).

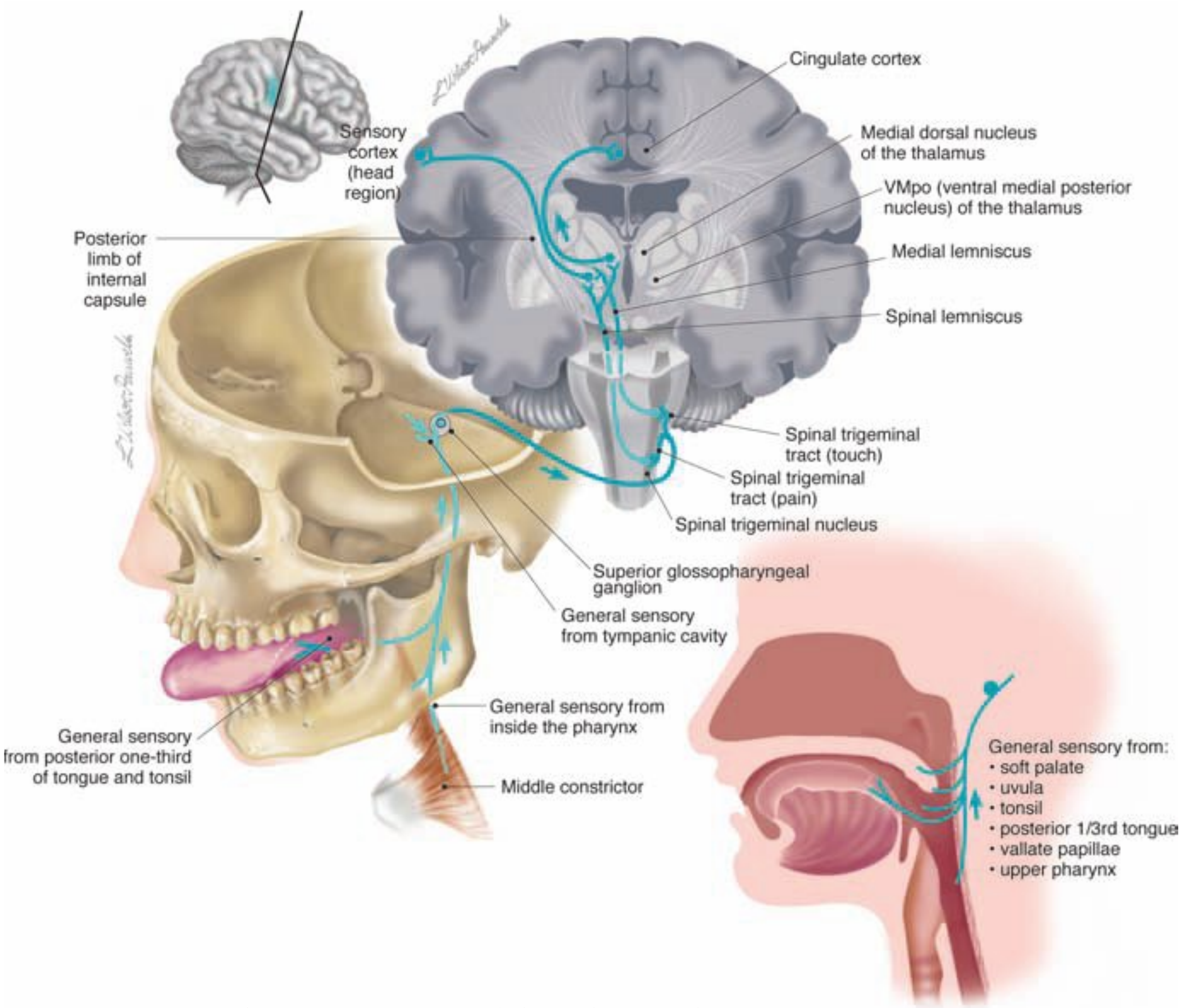
Cingulate cortex
Sensory cortex (head region)
Medial dorsal nucleus of the thalamus
VMpo (ventral medial posterior nucleus) of the thalamus
Posterior limb of internal capsule
Medial lemniscus
Spinal lemniscus
Spinal trigeminal tract (touch)
Spinal trigeminal tract (pain)
Spinal trigeminal nucleus
Superior glossopharyngeal ganglion
General sensory from tympanic cavity
General sensory from inside the pharynx
General sensory from posterior one-third of tongue and tonsil
Middle constrictor
General sensory from:
• soft palate
• uvula
• tonsil
• posterior 1/3rd tongue
• vallate papillae
• upper pharynx

FIGURE IX–3 General sensory component of glossopharyngeal nerve

medial nucleus of the thalamus, which project to the cingulate cortex where the emotional component of pain is mediated (see Fig. IX–3).

Glossopharyngeal axons carrying touch signals enter the medulla and synapse in the pontine trigeminal nucleus. Axons of secondary neurons cross the midline in the medulla and synapse in the VMpo of the contralateral thalamus. Axons of thalamic neurons project to the sensory cortex. These axons are the sensory limb of the "gag" reflex (see "Case History Guiding Questions," #7). These sensations operate at a "conscious" level of awareness.

VISCERAL SENSORY (AFFERENT) COMPONENT

Visceral sensory fibers operate at a "subconscious" level of awareness. Chemoreceptors in the carotid body monitor oxygen (O_2), carbon dioxide (CO_2), and acidity/

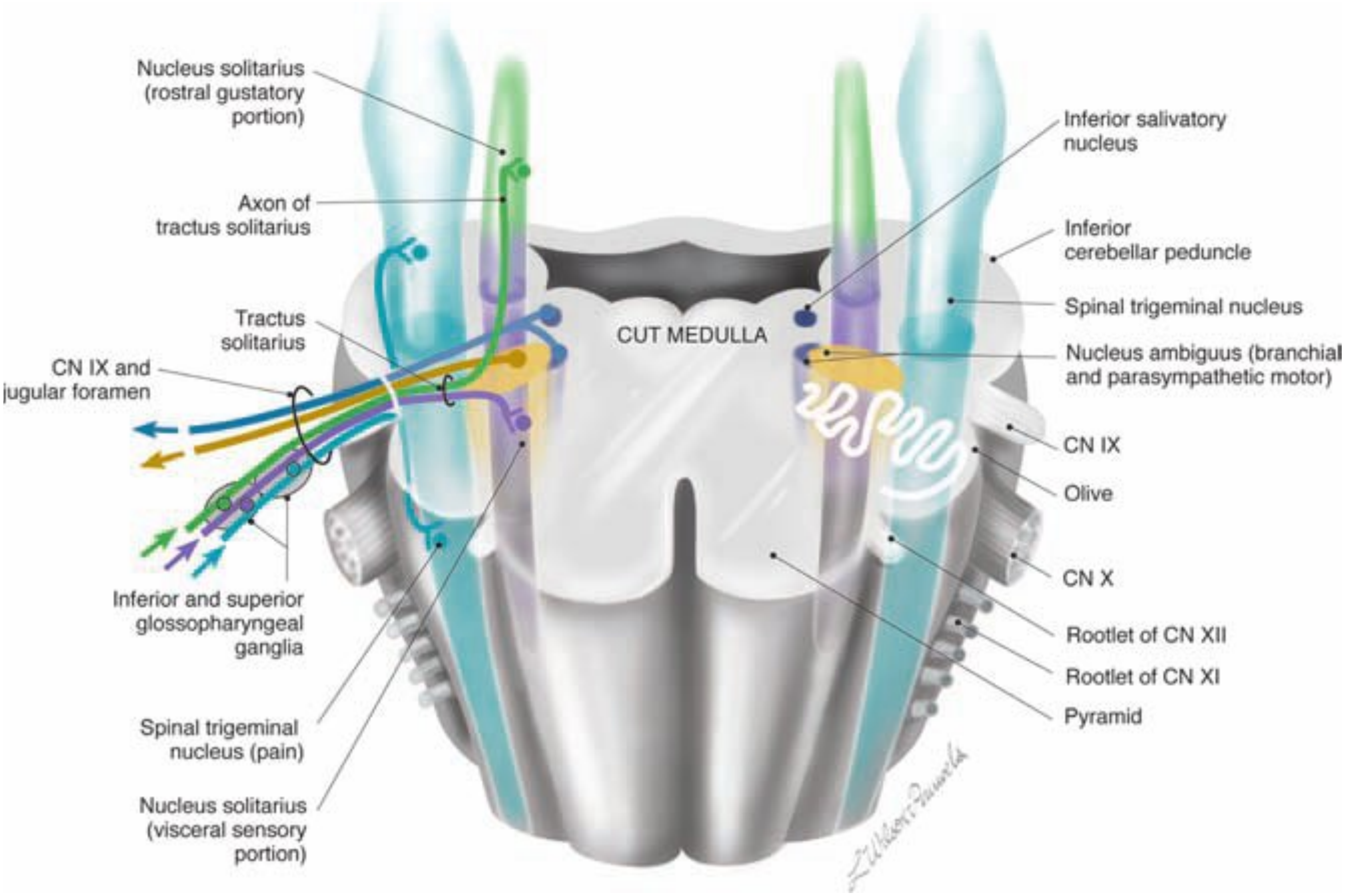
Nucleus solitarius (rostral gustatory portion)
Axon of tractus solitarius
Tractus solitarius
CN IX and jugular foramen
Inferior and superior glossopharyngeal ganglia
Spinal trigeminal nucleus (pain)
Nucleus solitarius (visceral sensory portion)
CUT MEDULLA
Inferior salivatory nucleus
Inferior cerebellar peduncle
Spinal trigeminal nucleus
Nucleus ambiguus (branchial and parasympathetic motor)
CN IX
Olive
CN X
Rootlet of CN XII
Rootlet of CN XI
Pyramid

FIGURE IX–4 Cross-section of the medulla at the point of entry of cranial nerve IX illustrating the nuclei associated with this nerve.

alkalinity (pH) levels in circulating blood, and baroreceptor (stretch receptor) nerve endings in the walls of the carotid sinus monitor arterial blood pressure (Fig. IX–5).

Visceral sensations from the walls of the carotid body and sinus ascend in the carotid nerve (Fig. IX–6; see also Fig. IX–5), in close proximity to the carotid artery, to reach the inferior glossopharyngeal ganglion where the nerve cell bodies are located. Central processes of the ganglionic neurons enter the medulla and descend in the tractus solitarius to synapse with nucleus solitarius neurons in the middle third of the nucleus (see Fig. IX–4). From this nucleus, connections are made with the reticular formation and the hypothalamus for the appropriate reflex responses for the control of respiration, blood pressure, and cardiac output.

Carotid Body

The carotid body is a small (3 mm × 6 mm in diameter) chemoreceptor organ located at the bifurcation of the common carotid artery (see Figs. IX–5 and IX–6). Very low levels of O_2, high levels of CO_2, and lowered pH levels (increased acidity) in the blood all cause an increase in the frequency of sensory signal traffic in the carotid branch of the glossopharyngeal nerve.

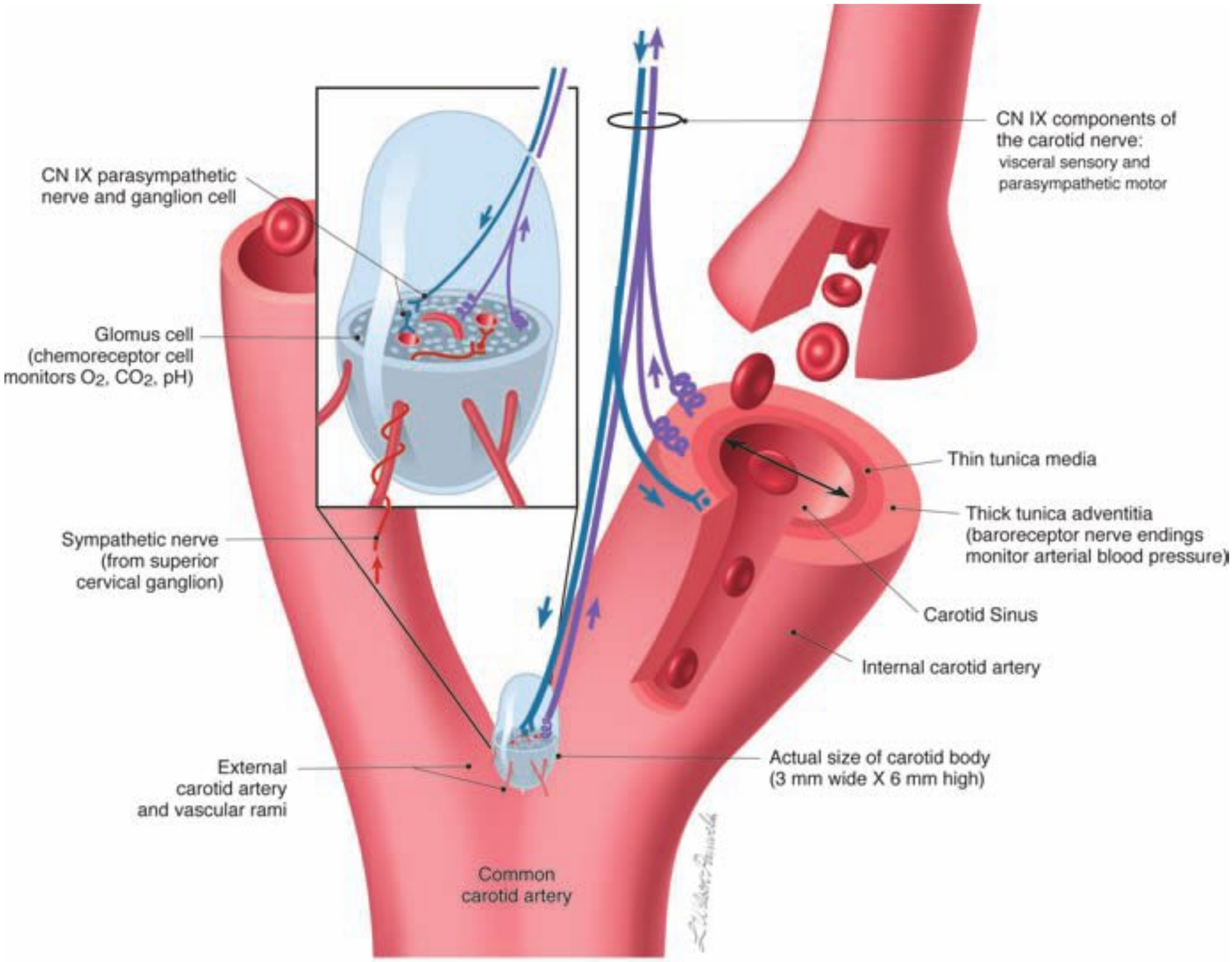
CN IX parasympathetic nerve and ganglion cell
Glomus cell (chemoreceptor cell monitors O2, CO2, pH)
Sympathetic nerve (from superior cervical ganglion)
External carotid artery and vascular rami
Common carotid artery
CN IX components of the carotid nerve: visceral sensory and parasympathetic motor
Thin tunica media
Thick tunica adventitia (baroreceptor nerve endings monitor arterial blood pressure)
Carotid Sinus
Internal carotid artery
Actual size of carotid body (3 mm wide X 6 mm high)

FIGURE IX–5 The bifurcation of the common carotid artery demonstrating CN IX sensory signals from baroreceptors in the wall of the carotid sinus and chemoreceptors within the carotid body as well as parasympathetic efferents that are vasodilatory to vessels of the carotid body and sinus. (The carotid body is enlarged to show structural detail.)

The transduction mechanism is not well understood. Glomus type I cells within the carotid body produce neurotransmitters that can excite or inhibit sensory nerve endings in response to changing conditions in the blood. In return, the nerve endings may alter the function of the glomus cells, possibly in a way that changes their sensitivity to O_2, CO_2, and pH.

Carotid Sinus

The carotid sinus is a dilation of the internal carotid artery at its origin from the common carotid artery. It responds to changes in arterial blood pressure. The adventitia contains many sensory nerve endings (stretch receptors) of the

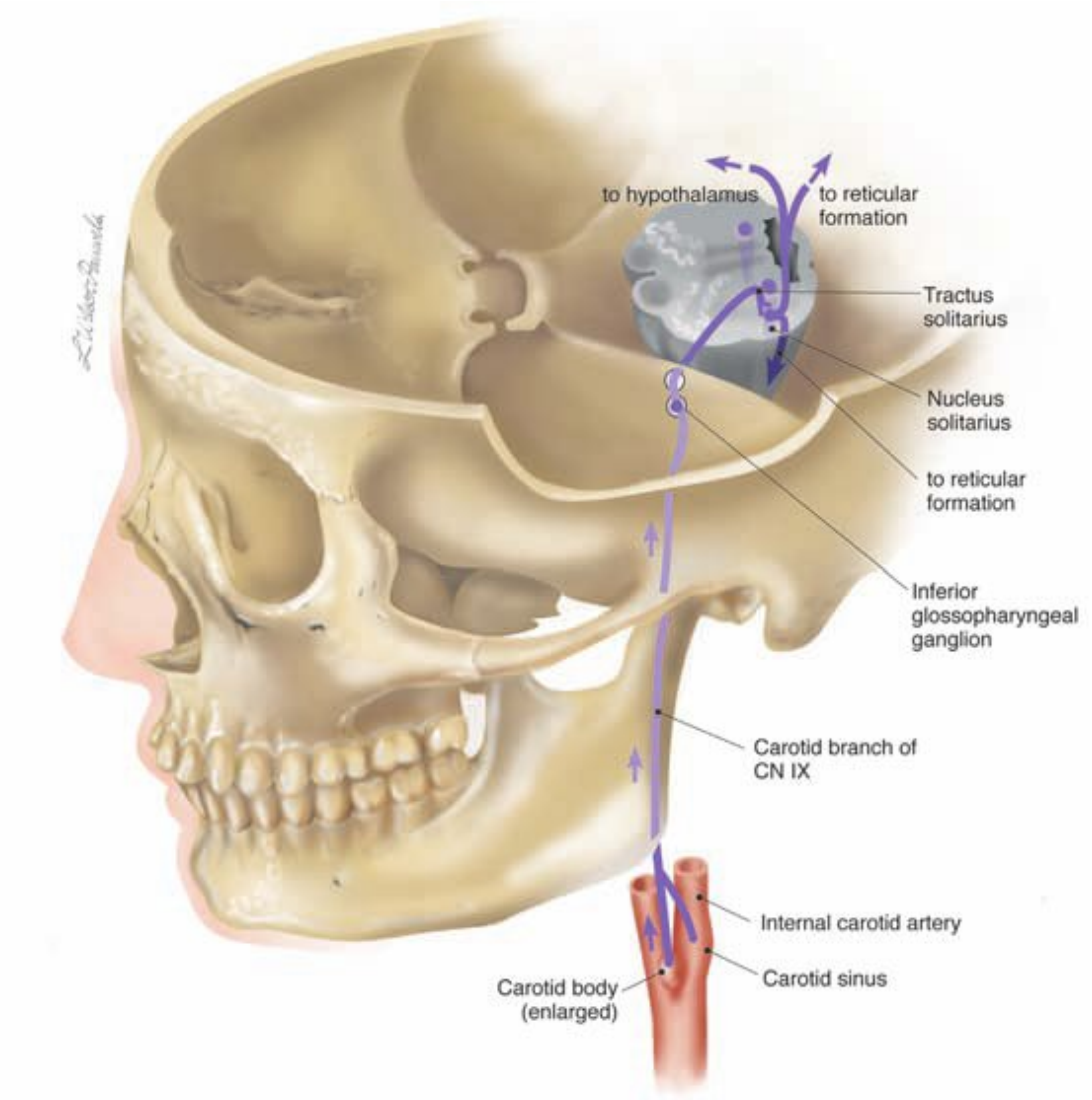

FIGURE IX–6 Visceral sensory component of the glossopharyngeal nerve–elevated brain stem.

glossopharyngeal nerve that monitor stretching of the sinus walls in response to increases in blood pressure within the sinus, thereby initiating impulses that reflexively lower the pressure (see Figs. IX–5 and IX–6). Because the internal carotid artery is the direct arterial flow to the brain, it is essential that blood pressure increases be monitored constantly and adjusted accordingly

SPECIAL SENSORY (AFFERENT) COMPONENT

Taste buds on the posterior one-third of the tongue are associated predominantly with vallate and foliate papillae (see “Facial Nerve VII” for a description of taste). Sensory processes from the taste buds travel in the glossopharygeal nerve toward their cell bodies in the inferior glossopharyngeal ganglion. Central processes from

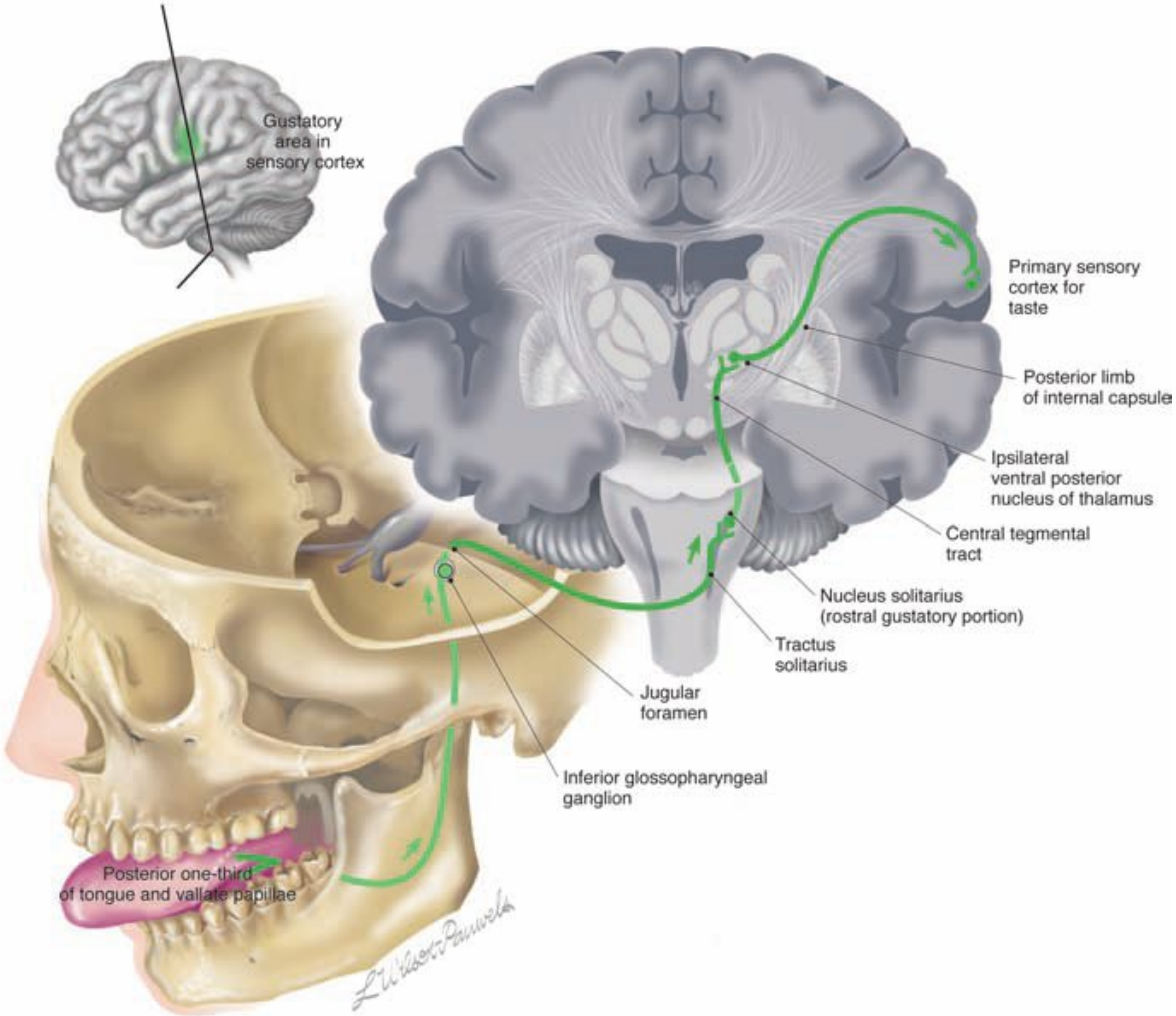
Gustatory area in sensory cortex
Primary sensory cortex for taste
Posterior limb of internal capsule
Ipsilateral ventral posterior nucleus of thalamus
Central tegmental tract
Nucleus solitarius (rostral gustatory portion)
Tractus solitarius
Jugular foramen
Inferior glossopharyngeal ganglion
Posterior one-third of tongue and vallate papillae
L Wilson-Pauwels

FIGURE IX–7 Special sensory component (for taste) of the glossopharyngeal nerve.

these neurons pass through the jugular foramen, enter the medulla, and ascend in the tractus solitarius to synapse in the nucleus solitarius (rostral gustatory portion) (Fig. IX–7; see also Fig. IX–4). Axons of cells in the nucleus solitarius then ascend in the central tegmental tract of the brain stem to reach the ipsilateral* ventral posterior nucleus of the thalamus. From the thalamus, fibers ascend through the posterior limb of the internal capsule to reach the primary sensory cortex in the inferior third of the postcentral gyrus and adjacent surface of the insula where taste is perceived.

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*Unlike the more recently evolved somatosensory system, chemosensation, that is, olfaction (CN I) and taste (CNs VII and IX), are perceived in the ipsilateral cortex.

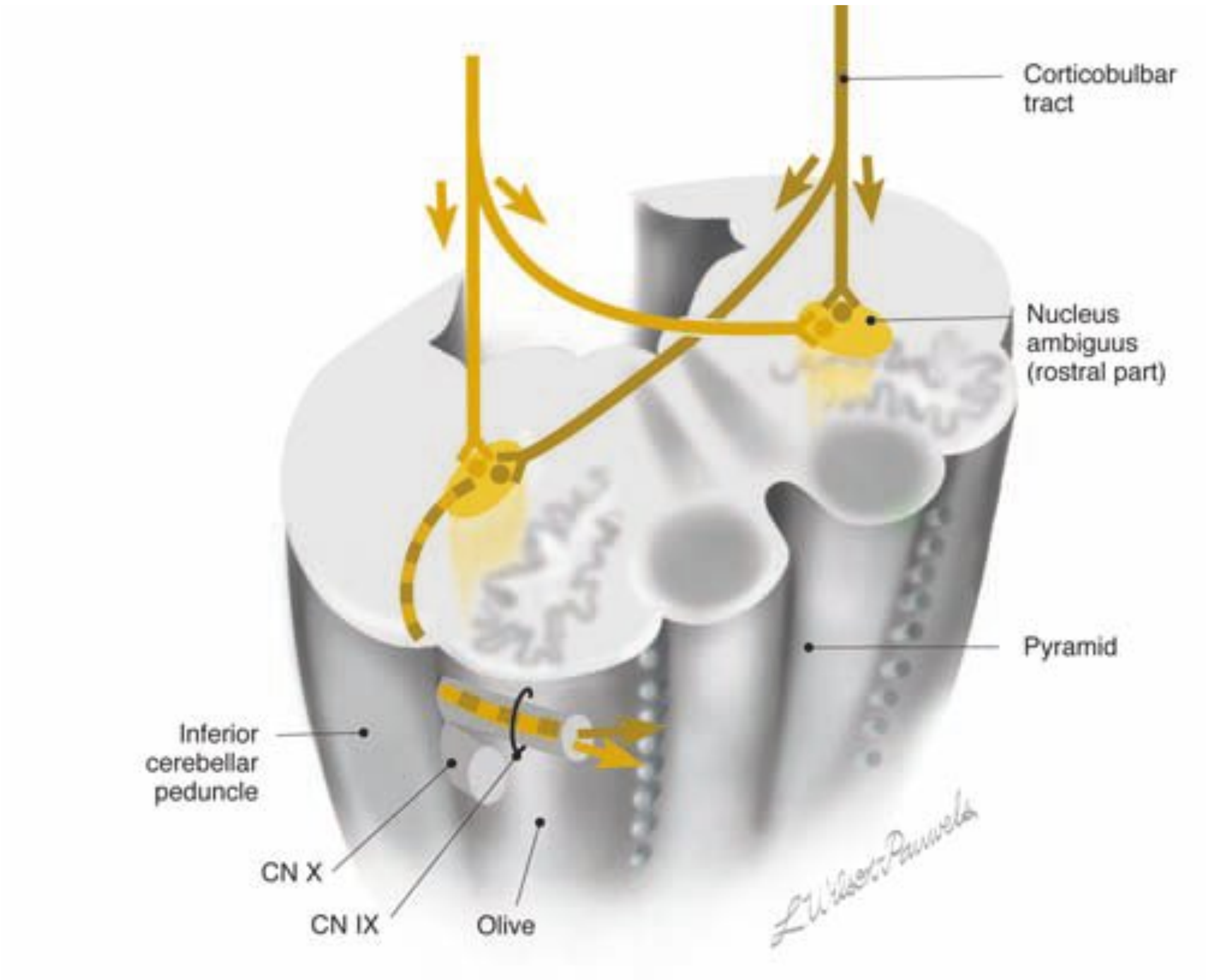

FIGURE IX–8 Branchial motor component of the glossopharyngeal nerve (section through the rostral part of the medulla).

BRANCHIAL MOTOR (EFFERENT) COMPONENT

In response to information received from the premotor sensory association cortex and other cortical areas, upper motor neurons in the primary motor cortex send impulses via corticobulbar tracts through the internal capsule and the basis pedunculi to synapse bilaterally on the lower motor neurons in the rostral part of the nucleus ambiguus (Figs. IX–8 and IX–9). The axons of these lower motor neurons join the other modalities of CN IX to leave the cranial cavity through the jugular foramen just rostral to the vagus and accessory nerves. The branchial motor axons branch off as a muscular branch that descends in the neck deep to the styloid process of the sphenoid bone and anterior to the internal carotid artery. The motor branch curves forward around the posterior border of the stylopharyngeus muscle and enters it to innervate its muscle fibers. The stylopharyngeus muscle elevates the pharynx during swallowing and speech (see Fig. IX–9).

PARASYMPATHETIC MOTOR (VISCERAL EFFERENT) COMPONENT

Preganglionic neurons of the parasympathetic motor fibers are located in both the inferior salivatory nucleus (see Fig. IX–4) that sends secretomotor input to the

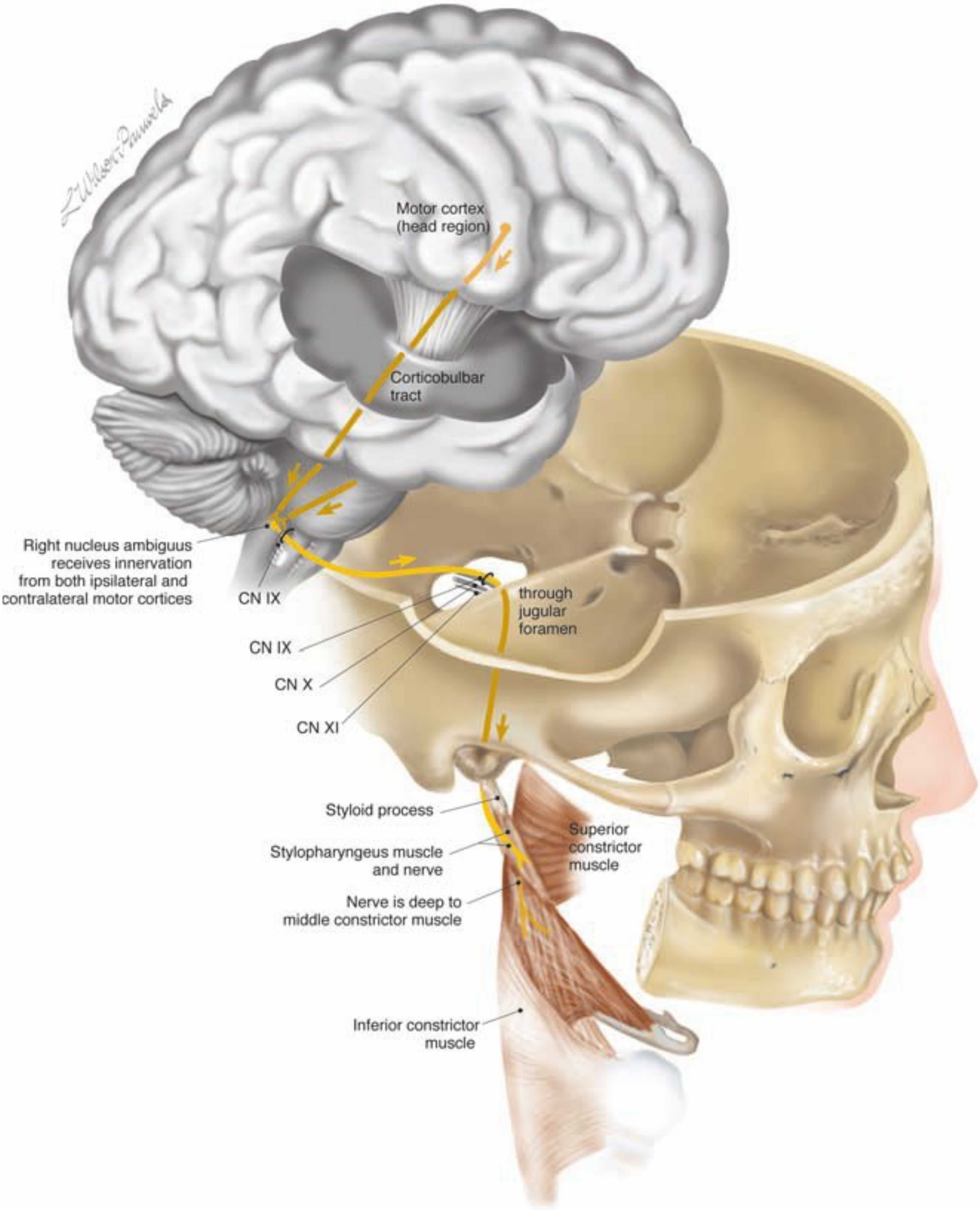
Motor cortex
(head region)
Corticobulbar
tract
Right nucleus ambiguus receives innervation from both ipsilateral and contralateral motor cortices
CN IX
through jugular foramen
CN IX
CN X
CN XI
Styloid process
Stylopharyngeus muscle and nerve
Superior constrictor muscle
Nerve is deep to middle constrictor muscle
Inferior constrictor muscle
L Wilson-Pauwels

FIGURE IX–9 Branchial motor component of the glossopharyngeal nerve.

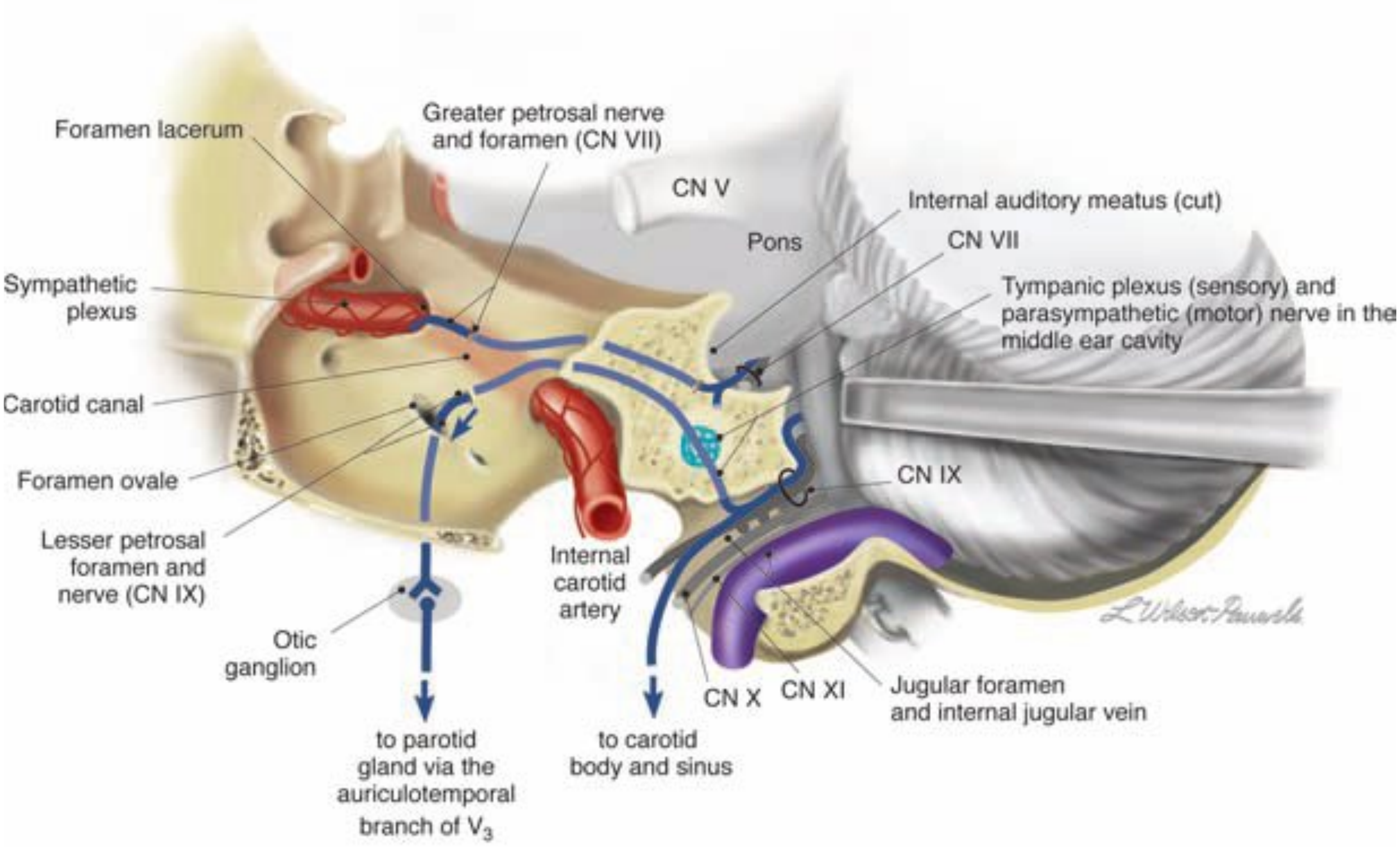
Foramen lacerum
Greater petrosal nerve and foramen (CN VII)
CN V
Internal auditory meatus (cut)
Pons
CN VII
Sympathetic plexus
Tympanic plexus (sensory) and parasympathetic (motor) nerve in the middle ear cavity
Carotid canal
Foramen ovale
CN IX
Lesser petrosal foramen and nerve (CN IX)
Internal carotid artery
Otic ganglion
CN X
CN XI
Jugular foramen and internal jugular vein
to parotid gland via the auriculotemporal branch of V3
to carotid body and sinus

FIGURE IX–10 Lesser petrosal nerve (parasympathetic motor IX) and greater petrosal nerve (parasympathetic motor VII) and surrounding structures (sagittal section through the jugular foramen showing cut petrous temporal bone).

parotid gland and the nucleus ambiguus in the medulla that sends vasodilatory fibers to the carotid body and sinus.

The inferior salivatory nucleus is influenced by stimuli from the hypothalamus via the dorsal longitudinal fasciculus (e.g., dry mouth in response to fear) and the olfactory system (e.g., salivation in response to smelling food). Axons from the inferior salivatory nucleus follow a tortuous course to their target structure, the parotid gland. They join the other components of CN IX in the medulla and travel with them into the jugular foramen (Fig. IX–10; see also Fig. IX–4). At the glossopharyngeal ganglion, these parasympathetic motor fibers leave the other modalities of CN IX as a component of the tympanic branch. They ascend to traverse the tympanic canaliculus and enter the tympanic cavity. Here they pass along the tympanic plexus located on the surface of the promontory of the middle ear cavity. From the tympanic plexus, the parasympathetic motor fibers form the lesser petrosal nerve that goes through a small canal back into the cranium to reach the internal surface of the temporal bone in the middle cranial fossa. The nerve leaves the cranial cavity again through a small opening, the lesser petrosal foramen, to become the lesser petrosal nerve that then passes forward to descend through the foramen ovale to synapse in the otic ganglion (see Fig. IX–10). From the otic ganglion, postganglionic fibers join the auriculotemporal nerve (a branch

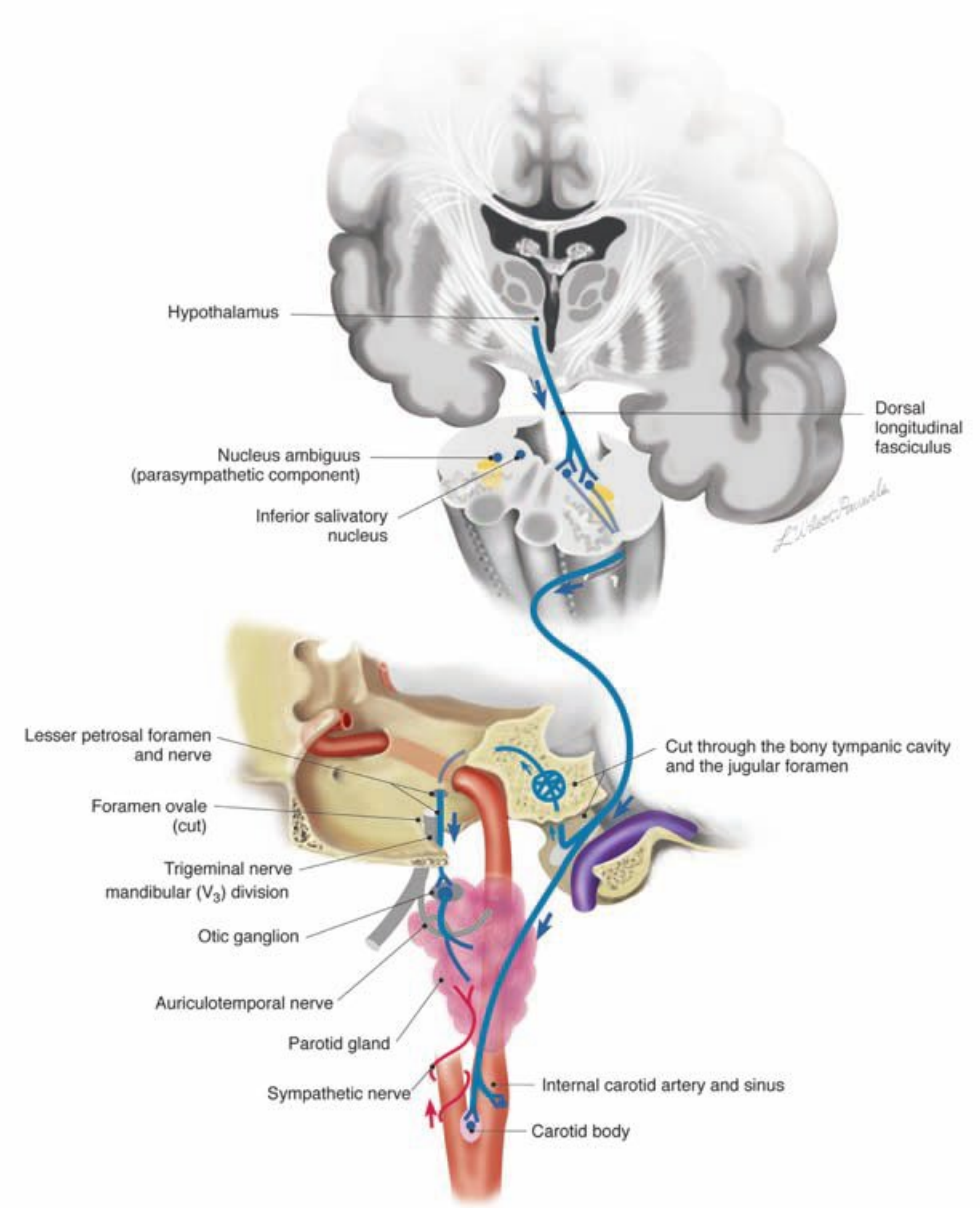
Hypothalamus
Dorsal longitudinal fasciculus
Nucleus ambiguus (parasympathetic component)
Inferior salivatory nucleus
L.Wilson-Pauwels
Lesser petrosal foramen and nerve
Foramen ovale (cut)
Trigeminal nerve mandibular (V_3) division
Otic ganglion
Auriculotemporal nerve
Parotid gland
Sympathetic nerve
Cut through the bony tympanic cavity and the jugular foramen
Internal carotid artery and sinus
Carotid body

FIGURE IX–11 Parasympathetic motor components of the glossopharyngeal nerve (CN IX) to: parotid gland, internal carotid artery and sinus, and carotid body.

of V_3) to supply secretomotor and vasodilatory input to the parotid gland (Fig. IX–11).[†]

The nucleus ambiguus is influenced by motor fibers from the hypothalamus and reticular formation. Parasympathetic axons from the nucleus ambiguus join the other components of CN IX in the medulla and travel with them through the jugular foramen. These axons travel with the carotid branch of CN IX to the carotid body and to the carotid sinus where their preganglionic axons synapse on parasympathetic ganglia within the carotid body and in the walls of the carotid sinus. Their role is vasodilation of the blood vessels within the carotid body as well as the carotid sinus.

The carotid body and sinus are also supplied by a plexus of sympathetic motor nerves traveling with the blood vessels surrounding the external carotid artery and its branches in the area (see Figs. IX–5 and IX–11). Their role in carotid body function is vasoconstriction of the blood vessels.

CASE HISTORY GUIDING QUESTIONS

1. What is glossopharyngeal neuralgia?
2. What is the cause of glossopharyngeal neuralgia?
3. What is allodynia?
4. Why did swallowing cause Allen pain?
5. Why did Allen collapse to the floor?
6. Why was carbamazepine used to treat Allen's symptoms?
7. What is the gag reflex, and why don't we gag every time a bolus of food passes through the pharynx?

1. What is glossopharyngeal neuralgia[‡]?

Glossopharyngeal neuralgia is characterized by severe, sharp, stabbing pain in the region of the tonsil, radiating to the ear. It is similar to trigeminal neuralgia (see Chapter V) in its timing and its ability to be triggered by various stimuli. For example, pain may be initiated by yawning, swallowing, or contact with food in the tonsilar region. In rare instances, the pain may be associated with syncope (a fall in heart rate and blood pressure resulting in fainting).

[†] Under stimulation, glands increase their secretion by three to four orders of magnitude above the basal level and they reguire a robust blood flow to provide the raw materials. This is driven by parasympathetic stimulation.

[‡] Some authors use the term "vagoglossopharyngeal neuralgia" or "glossopharyngeal and vagal neuralgia" instead of "glossopharyngeal neuralgia," implying that the pain can radiate into the distribution of the vagus nerve as well as that of the glossopharyngeal nerve. However, the original term, "glossopharyngeal neuralgia," is recognized by most neurologists and is more commonly used.

2. What is the cause of glossopharyngeal neuralgia?

Typically, glossopharyngeal neuralgia is idiopathic; that is, no cause can be identified. Occasionally, glossopharyngeal neuralgia is secondary to the compression of CN IX by carotid aneurysms, oropharyngeal malignancies, peritonsillar infections, or lesions at the base of the skull.

3. What is allodynia?

Allodynia is pain that results from a touch stimulus that normally would not cause pain. The pain is usually burning or stabbing in quality.

4. Why did swallowing cause Allen pain?

Sensory nerve endings in the mucosa of Allen's pharynx were stimulated by the passage of a bolus of food and by the movements of the underlying muscles involved in swallowing, coughing, and speaking. These usually harmless stimuli set off a barrage of pain impulses within the nervous system.

5. Why did Allen collapse to the floor?

Allen collapsed on the floor because his blood pressure dropped so low that he could not maintain adequate blood flow to his brain. The mechanism for Allen's fall in blood pressure and heart rate is not well understood. One hypothesis is that when the pain is most intense, general sensory impulses from the pharynx stimulate the nucleus of the tractus solitarius, which, in turn, stimulates vagal nuclei. Impulses from the parasympathetic component of the vagus nerve act to slow the heart rate and decrease blood pressure.

Alternatively, the rapidly firing general sensory afferents may, through ephaptic transmission,[§] give rise to action potentials in the visceral sensory axons from the carotid sinus as they travel together in the main trunk of the glossopharyngeal nerve. The carotid nerve would then falsely report increased blood pressure, causing a reflexive decrease in heart rate and blood pressure via connections in the brain stem.

6. Why was carbamazepine used to treat Allen's symptoms?

Carbamazepine is a sodium channel blocking drug that is used to treat a wide range of conditions from seizures to neuropathic pain. Carbamazepine reduces the glossopharyngeal nerve's ability to fire trains of action potentials at high frequency and, therefore, shortens the duration of the paroxysm and frequently abolishes the attacks.

7. What is the gag reflex, and why don't we gag every time a bolus of food passes through the pharynx?

The gag is a protective reflex that prevents the entry of foreign objects into the alimentary and respiratory passages. A touch stimulus to the back of the tongue or pharyngeal walls elicits the gag (see "Clinical Testing," later). Because the passage of a bolus of food through the mouth and pharynx stimulates the tongue and pharyngeal walls, we have to ask why swallowing does not elicit a gag reflex.

[§] Ephaptic transmission occurs when a rapidly firing nerve changes the ionic environment of adjacent nerves sufficiently to give rise to action potentials in them. These "artificial synapses" often result from a breakdown in myelin.

Although the exact mechanism is not well understood, it is generally accepted that when a swallowing sequence is initiated, probably by signals from the cerebral cortex to swallowing center(s) in the brain stem, simultaneous inhibitory signals are sent to the gag center in the brain stem to turn off the gag reflex. A gag is not a partial vomit, but it can include vomiting.

Four events occur in the gag reflex (Fig. IX–12):

1. An irritant is sensed by the tongue (CNs VII and IX).
2. The soft palate elevates and is held firmly against the posterior pharyngeal wall, closing off the upper respiratory airway (CN X).
3. The glottis is closed to protect the lower respiratory passages (CN X).

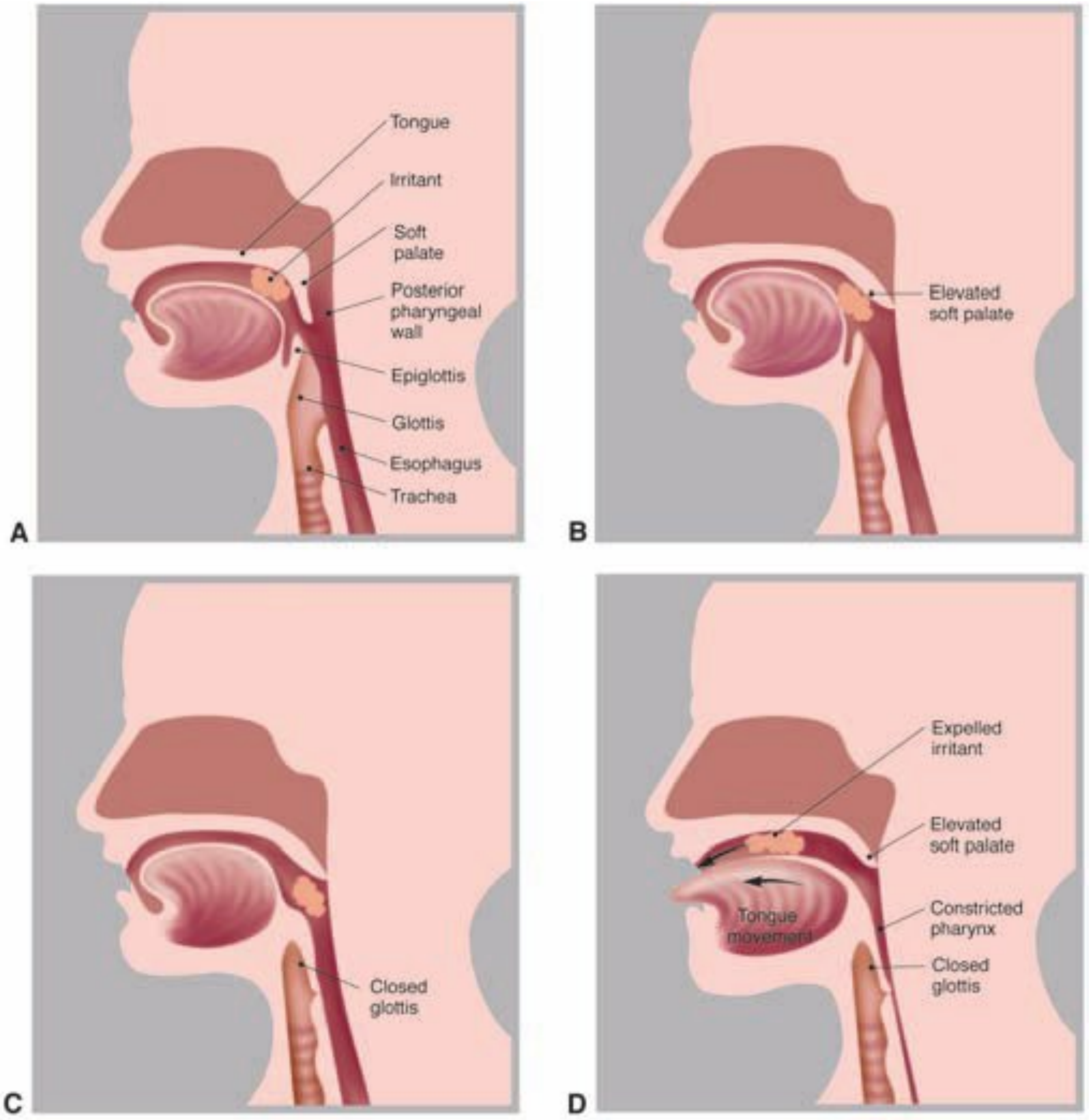

FIGURE IX–12 Steps in the gag reflex. **A**, Irritant in the mouth. **B**, Soft palate elevates, closing the upper respiratory airway. **C**, Glottis is closed to protect the lower respiratory airway. **D**, Pharyngeal wall constricts causing expulsion of the foreign object.

4. The pharynx is constricted to prevent entry into the alimentary tract, the pharyngeal wall constricts (CN X) and the tongue moves the contents of the pharynx forward to expel the offending foreign object from the mouth (CN XII).

The sensitivity of the gag reflex can be affected by cortical events. It can be suppressed almost completely or enhanced such that even brushing the teeth or the sight of a dental impression tray can lead to gagging.

CLINICAL TESTING

Although general, visceral, and special sensory, as well as branchial and parasympathetic motor fibers, make up CN IX, from a practical point of view, only the general sensory component is tested at the bedside. Clinically, CNs IX and X are assessed by testing the gag reflex. The gag reflex involves both CNs—the glossopharyngeal being the sensory (afferent) input and the vagus carrying the motor (efferent) components. When assessing the gag reflex, right and left sides of the upper pharynx should be lightly touched with a tongue depressor (Fig. IX–13). The sensory limb of the reflex is considered to be intact if the pharyngeal wall can be seen to contract after each side has been touched.

Figure IX–14 is a schematic representation of all the CNs involved in the gag reflex. As well as depicting the role of CN IX and CN X, described earlier, the illus-

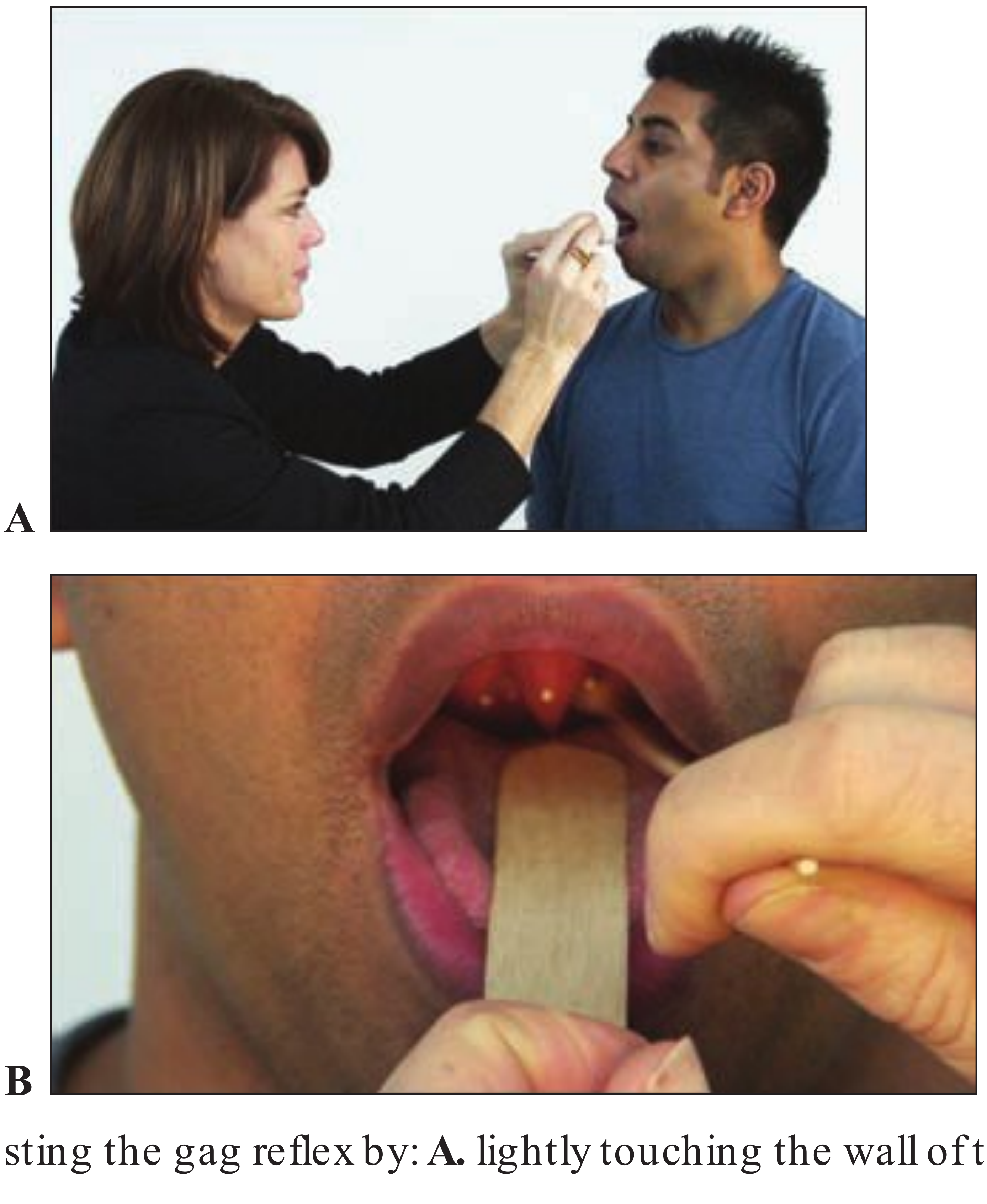

FIGURE IX–13 Testing the gag reflex by: **A.** lightly touching the wall of the pharynx, **B.** close up.

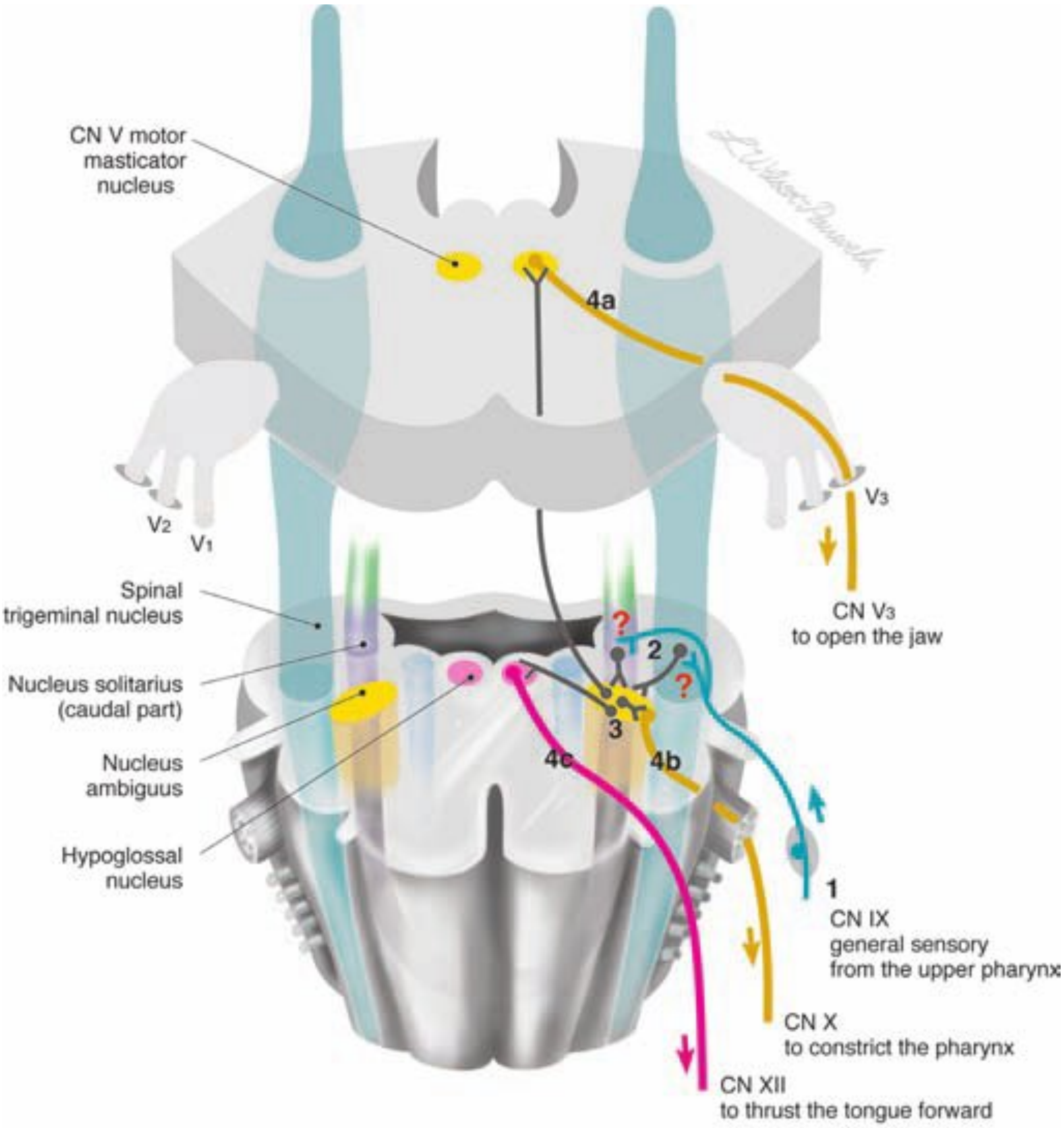
CN V motor masticator nucleus
4a
V2
V1
V3
Spinal trigeminal nucleus
Nucleus solitarius (caudal part)
Nucleus ambiguus
Hypoglossal nucleus
?
2
?
3
4c
4b
CN V3 to open the jaw
1
CN IX general sensory from the upper pharynx
CN X to constrict the pharynx
CN XII to thrust the tongue forward

FIGURE IX–14 Gag reflex pathways.

1. Stimulation of the upper pharynx activates CN IX sensory neurons.
2. Interneurons (grey) in the spinal trigeminal nucleus or in the caudal part of nucleus solitarius send signals to
3. Nucleus ambiguus that, in turn, activate:
4. a. neurons in the masticator nucleus that open the jaw via CN V_3,
   b. other neurons in nucleus ambiguus that cause pharyngeal constriction via CN X and
   c. neurons in the hypoglossal nucleus that cause thrusting of the tongue via CN XII.

NB. The pathways for the gag reflex in the human are not known with certainty. Since the pharynx is a point of transition between the body wall and the viscera, sensory afferents may be somatic, visceral or both. The afferent limb of the gag reflex, therefore, may synapse in the spinal trigeminal nucleus, the nucleus solitarius, or both (indicated with red question marks).

tration highlights the role of CN V_3 in activating the mandibular depressor muscles and CN XII in tongue activation. (See also “Cranial Nerves Examination” on the website.)

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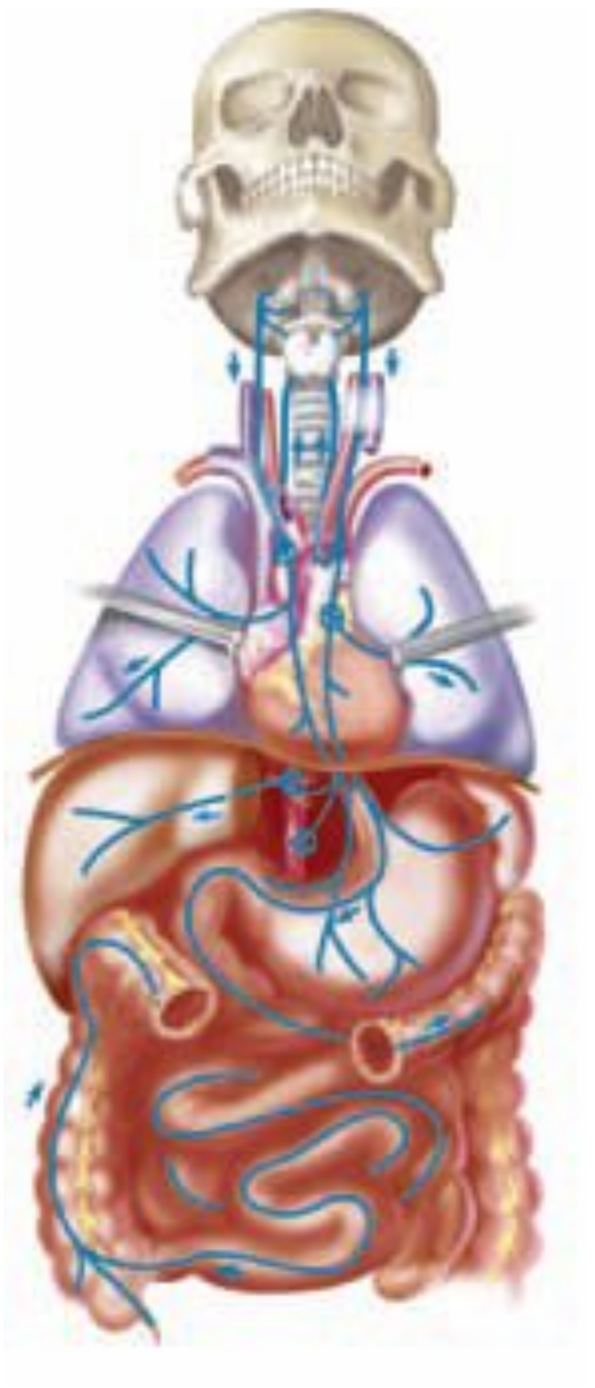

X

Vagus Nerve

CASE HISTORY

Ruth is a 46-year-old lawyer who, over the last few years, noticed a whooshing sound in her left ear when she lay on her left side at night. Ruth is a very busy woman with two teenage daughters and an active law practice, and because the noise did not keep her from sleeping at night, she paid little attention to it.

One afternoon, after playing a vigorous game of tennis with her daughter, she again became aware of the whooshing sensation in her left ear and noted that it seemed more intense with exercise. She had intended to see her family doctor but got distracted with an important defense and her daughter's upcoming graduation.

Over several months, Ruth noticed the whooshing sound was almost always present day or night, and she gradually developed problems with swallowing and a hoarse voice. Finally, Ruth made time to see her doctor.

On general examination, Ruth's doctor noted that she appeared fit and well. However, when the doctor placed a stethoscope on the base of her skull on the left side, he could hear a bruit (a whooshing sound). When he examined her cranial nerves, he found that she had an absent gag reflex on the left side and some weakness of her left sternomastoid* muscle. Ruth's doctor immediately referred her to a neurosurgeon who was concerned that this was a glomus tumor of the jugular foramen. He sent Ruth for magnetic resonance imaging (MRI) and an angiogram. The investigations confirmed the neurosurgeon's suspicions and a diagnosis of a glomus jugulare tumor was made. Ruth was subsequently scheduled for surgery to have the tumor removed.

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*"Sternomastoid" is a shortened form of "sternocleidomastoid" and is used in this text.

ANATOMY OF THE VAGUS NERVE

Vagus comes from the Latin word meaning "wandering." The vagus nerve "wanders" from the brain stem to reach almost to the splenic flexure of the colon. The vagus nerve is the largest visceral sensory (afferent) nerve of the cranial nerves. In addition, it has a relatively large parasympathetic motor component and a general sensory and branchial motor component (Fig. X–1). In the medulla, the vagal fibers are connected to four nuclei: the spinal nucleus of the trigeminal nerve (general sensory); the caudal portion of the nucleus of the tractus solitarius (visceral sensory); the nucleus ambiguus (branchial motor); and the dorsal vagal motor nucleus (parasympathetic visceral motor) (Fig. X–2 and Table X–1).

Course of the Vagus Nerve

The vagus nerve emerges from the medulla of the brain stem dorsal to the olive as 8 to 10 rootlets caudal to those of cranial nerve (CN) IX (see Fig. X–2). These rootlets converge into a flat cord that exits the skull through the jugular foramen. Two sensory ganglia, the superior (jugular) and the inferior (nodosum), are located on the vagus nerve. The superior ganglion is located within the jugular fossa of the petrous temporal bone, which together with the occipital bone, forms the jugular foramen. Within the jugular foramen, the vagus nerve is in close proximity to the jugular bulb, a swelling of the proximal part of the internal jugular vein containing the jugular glomus within its adventitia (see "Case History Guiding Questions," #1). The glomus jugulare, or tympanic body, is a collection of neuron-like cells that monitor blood oxygen (O_2), carbon dioxide (CO_2), and acidity/alkalinity (pH) levels. It is similar to the carotid body (see Chapter IX). After exiting the jugular foramen, the vagus nerve enlarges into a second swelling, the inferior ganglion (see Fig. X–2).

As the vagus nerve emerges through the jugular foramen, it lies within the same dural sheath as the accessory nerve (CN XI; Fig. X–3). For a short distance, the caudal branchial motor fibers of CN X travel with CN XI.[†] Just beyond the inferior ganglion, all the branchial motor fibers of cranial nerve X rejoin (see Fig. X–3).

In the neck, the vagus nerve lies posterior to and in a groove between the internal jugular vein and the internal carotid artery. It descends vertically within the carotid sheath (Fig. X–4), giving off branches to the pharynx, larynx, and constrictor muscles (Table X–2). The right recurrent laryngeal nerve branches from the right vagus nerve in the neck. Running anterior to the right subclavian artery, the nerve curves below and behind the artery to ascend posterior to it in a groove at the right side of the trachea behind the right common carotid artery. The left recurrent laryngeal nerve branches from the left vagus nerve in the thorax (see Fig. X–4).

[†] There is general lack of agreement whether these axons should be classified as the cranial root of the accessory nerve or the caudal part of the vagus nerve. In this text, we maintain the latter convention.

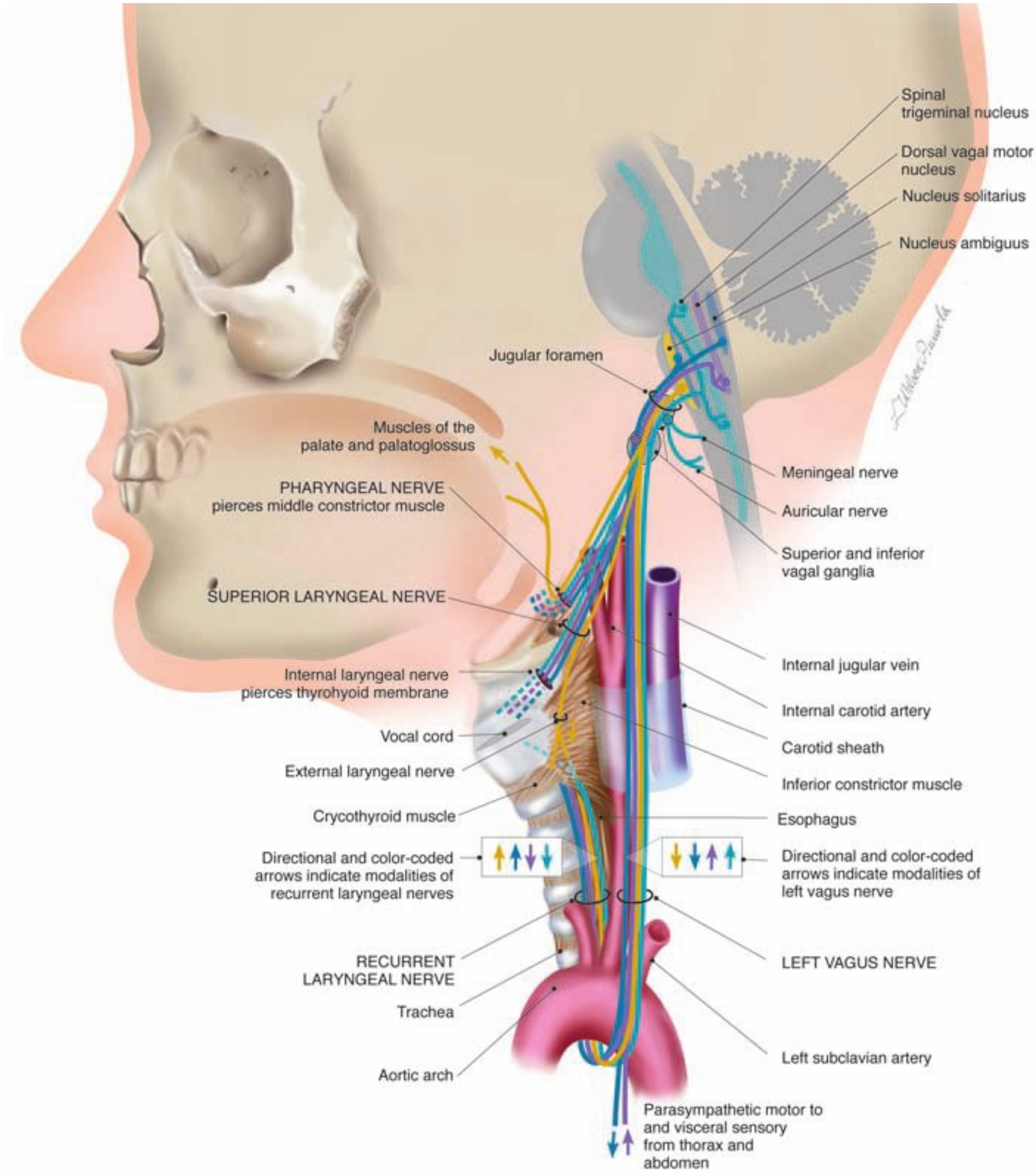

FIGURE X–1 Overview of the left vagus nerve.

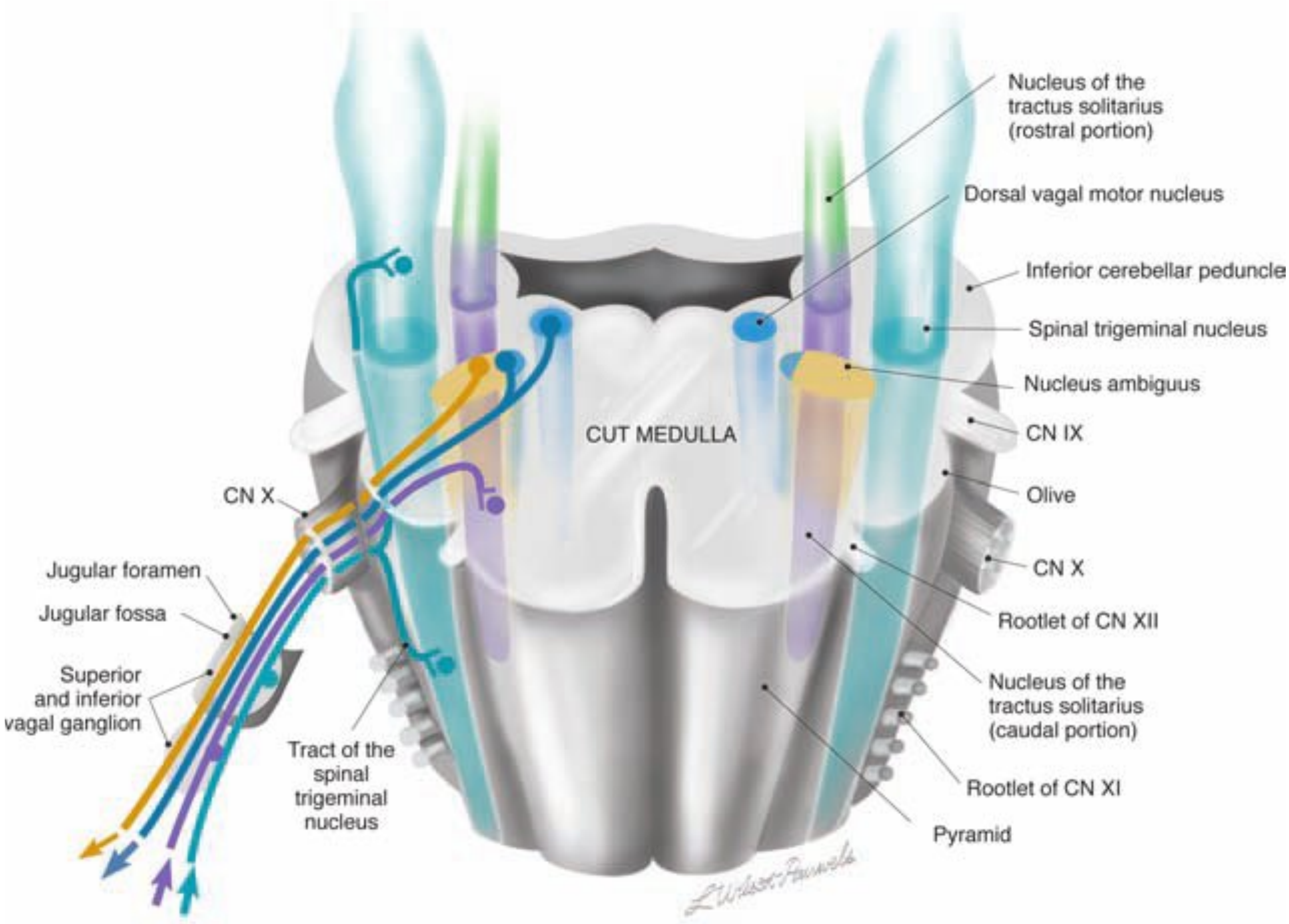
Nucleus of the tractus solitarius (rostral portion)
Dorsal vagal motor nucleus
Inferior cerebellar peduncle
Spinal trigeminal nucleus
Nucleus ambiguus
CN IX
Olive
CN X
Rootlet of CN XII
Nucleus of the tractus solitarius (caudal portion)
Rootlet of CN XI
Pyramid
CUT MEDULLA
CN X
Jugular foramen
Jugular fossa
Superior and inferior vagal ganglion
Tract of the spinal trigeminal nucleus

FIGURE X–2 Cross-section of the medulla at the point of entry of cranial nerve X illustrating the nuclei associated with this nerve.

TABLE X–1 Components, Nuclei, Ganglia and Functions of the Vagus Nerve (CN X)

Component*	Nucleus	Ganglion	Function
General sensory (afferent)	Spinal trigeminal nucleus	Superior vagal ganglion	For general sensation from the posterior meninges, concha, skin at the back of the ear and in the external acoustic meatus, part of the external surface of the tympanic membrane, the pharynx, and larnyx
Visceral sensory (afferent)	Nucleus of tractus solitarius	Inferior vagal ganglion	For visceral sensation from the lower pharynx, larynx, trachea, esophagus, and thoracic and abdominal viscera, stretch receptors in the walls of the aortic arch, and chemoreceptors in the aortic bodies adjacent to the arch
Branchial motor (efferent)	Nucleus Ambiguus		To innervate, via the pharyngeal plexus, the superior, middle, and inferior constrictors, levator palati, salpingopharyngeus, palatopharyngeus, and one muscle of the tongue, the palatoglossus, and to the intrinsic muscles of the larynx
Parasympathetic motor (visceral efferent)	Dorsal vagal motor nucleus Nucleus ambiguus		To innervate smooth muscle and stimulate glands of the pharynx, larynx, and thoracic and abdominal viscera To innervate cardiac muscle and the aortic bodies

*Some texts list special sense for taste as one of the components of this nerve. Because CN X carries so few taste fibers, this modality has been omitted.

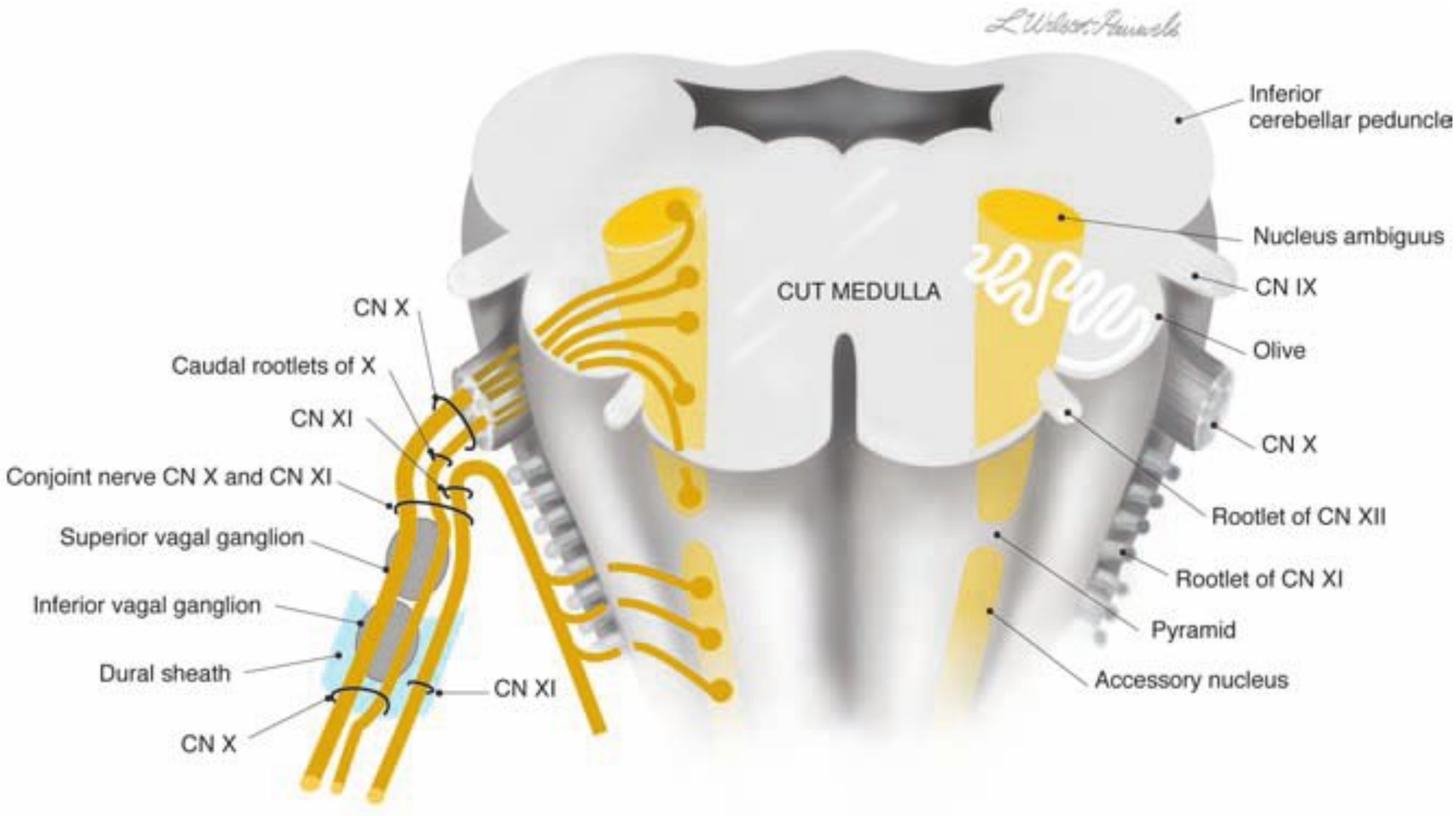

FIGURE X–3 Cross-section through the rostral (open) medulla demonstrating the branchial motor component of cranial nerve X including spinal rootlets of cranial nerve XI.

It curves below and behind the aortic arch to ascend at the left side of the trachea. From the root of the neck downward, the vagus nerve takes a different path on each side of the body to reach the cardiac, pulmonary, and esophageal plexuses. From the esophageal plexus, right and left gastric nerves emerge to supply the stomach and most of the abdominal viscera, ending close to the splenic flexure (see Table X–2).

GENERAL SENSORY (AFFERENT) COMPONENT

The general sensory component of CN X carries sensation (pain, touch, and temperature) from the

- Larynx.
- Lower part of pharynx.
- Concha and skin of the external ear and external acoustic meatus.
- External surface of the tympanic membrane.
- Meninges of the posterior cranial fossa.

Axons carrying general sensation from the vocal folds and the subglottis below the vocal folds accompany visceral sensory axons in the recurrent laryngeal nerves (Figs. X–5 and X–6). Similarly, axons carrying general sensation from the larynx above the vocal folds accompany visceral sensory axons in the internal laryngeal nerves. The internal laryngeal nerves leave the pharynx by piercing the thyrohyoid

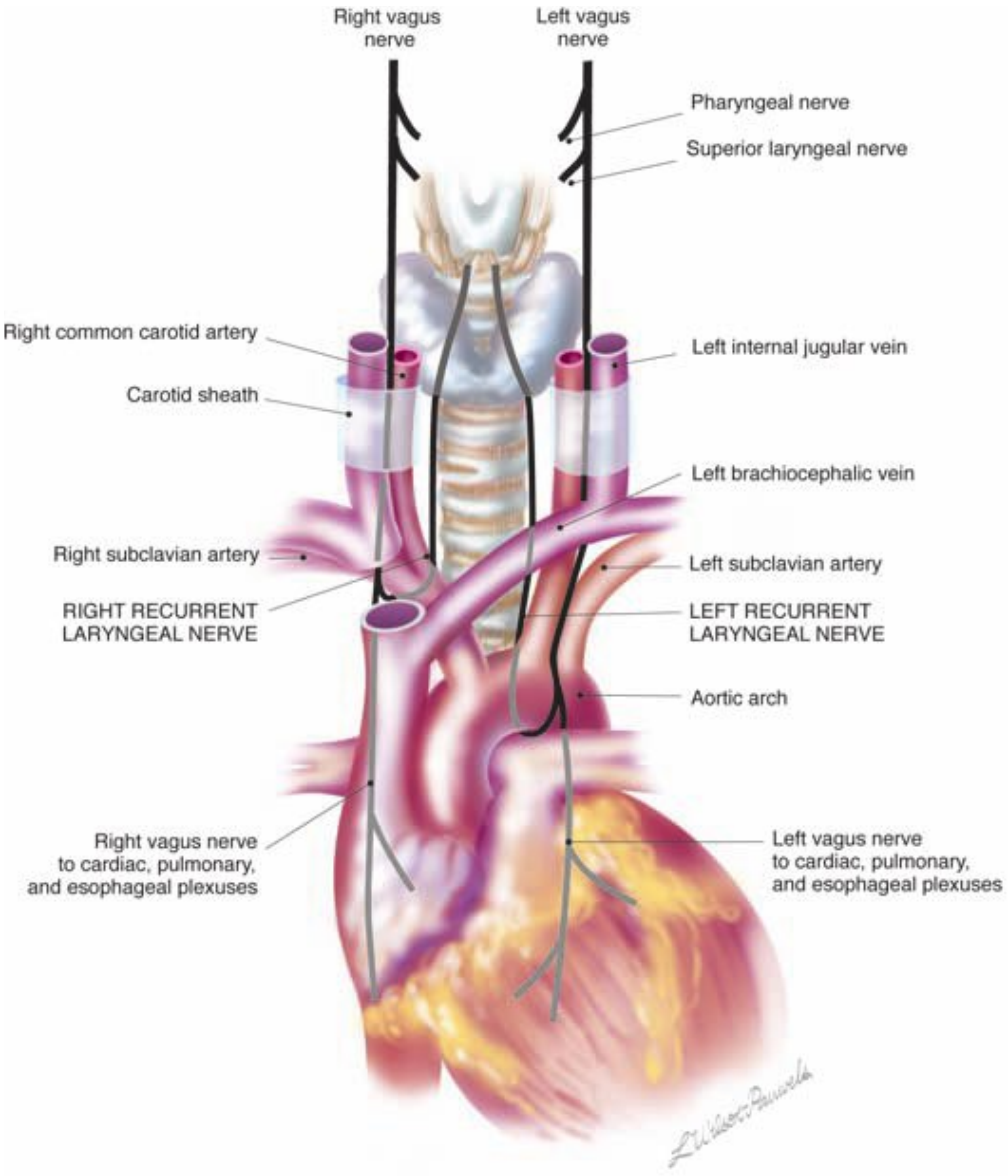
Right vagus nerve
Left vagus nerve
Pharyngeal nerve
Superior laryngeal nerve
Right common carotid artery
Left internal jugular vein
Carotid sheath
Left brachiocephalic vein
Right subclavian artery
Left subclavian artery
RIGHT RECURRENT LARYNGEAL NERVE
LEFT RECURRENT LARYNGEAL NERVE
Aortic arch
Right vagus nerve to cardiac, pulmonary, and esophageal plexuses
Left vagus nerve to cardiac, pulmonary, and esophageal plexuses

FIGURE X–4 Route of the right and left recurrent laryngeal nerves (nerve is shown in black for clarity).

membrane. They ascend in the neck uniting with the external laryngeal nerves (branchial motor) to form the superior laryngeal nerves (see Fig. X–1). General sensory fibers travel up the superior laryngeal nerves to join the rest of the vagus nerves and reach the inferior vagal ganglia.

General sensory fibers from the concha and the skin of the external ear, the external auditory canal, and the external surface of the tympanic membrane are carried in the auricular branch (see Fig. X–6). Stimulation of the auricular nerve of CN X in the external auditory meatus can cause reflex coughing, vomiting, and even fainting through reflex activation of the dorsal vagal motor nucleus. Sensory

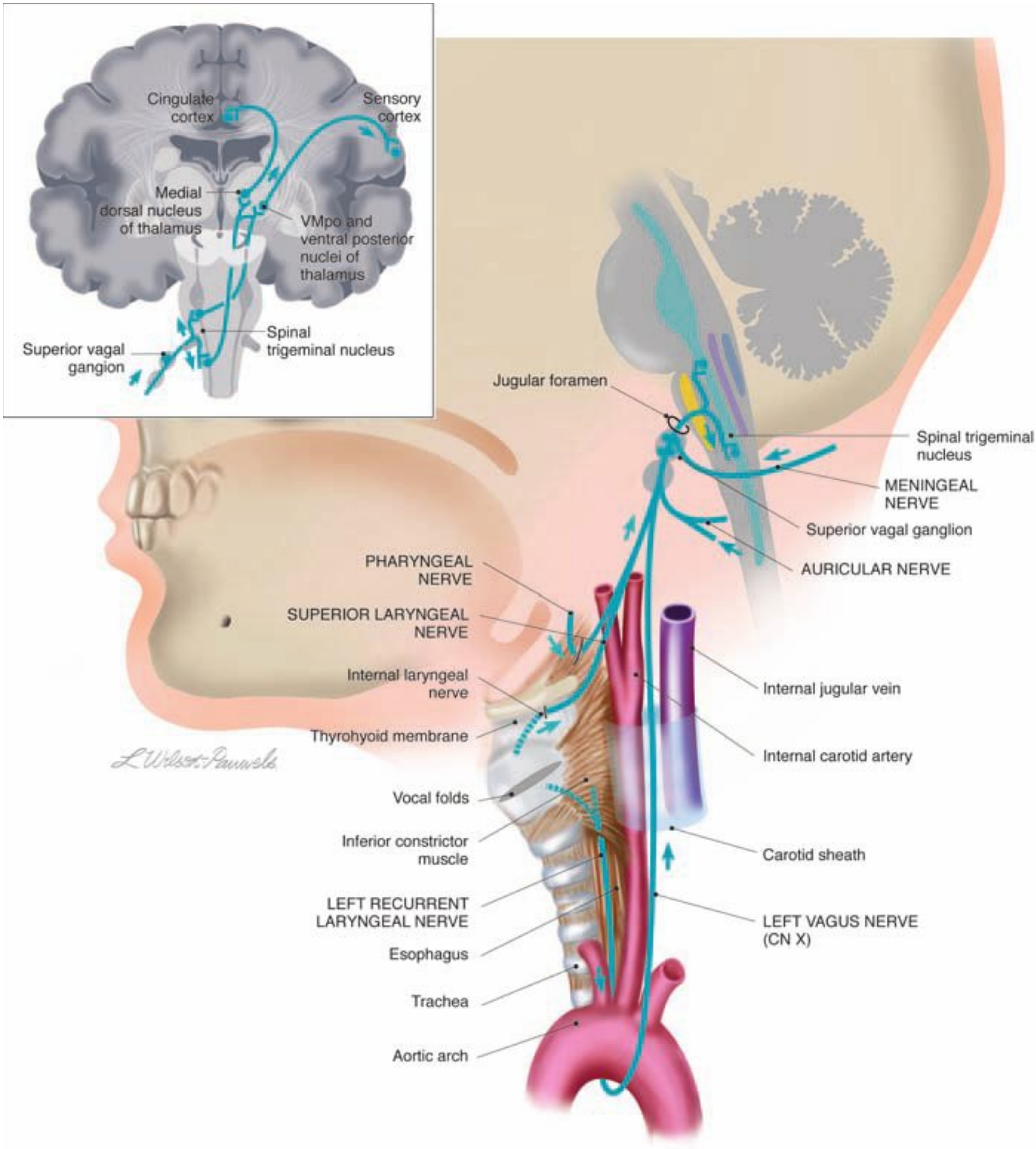

FIGURE X–5 General sensory component of the vagus nerve.

branches from the meninges of the posterior cranial fossa are carried in the meningeal nerve. The peripheral processes pass into the jugular fossa and enter the superior vagal ganglion, where their nerve cell bodies are located.

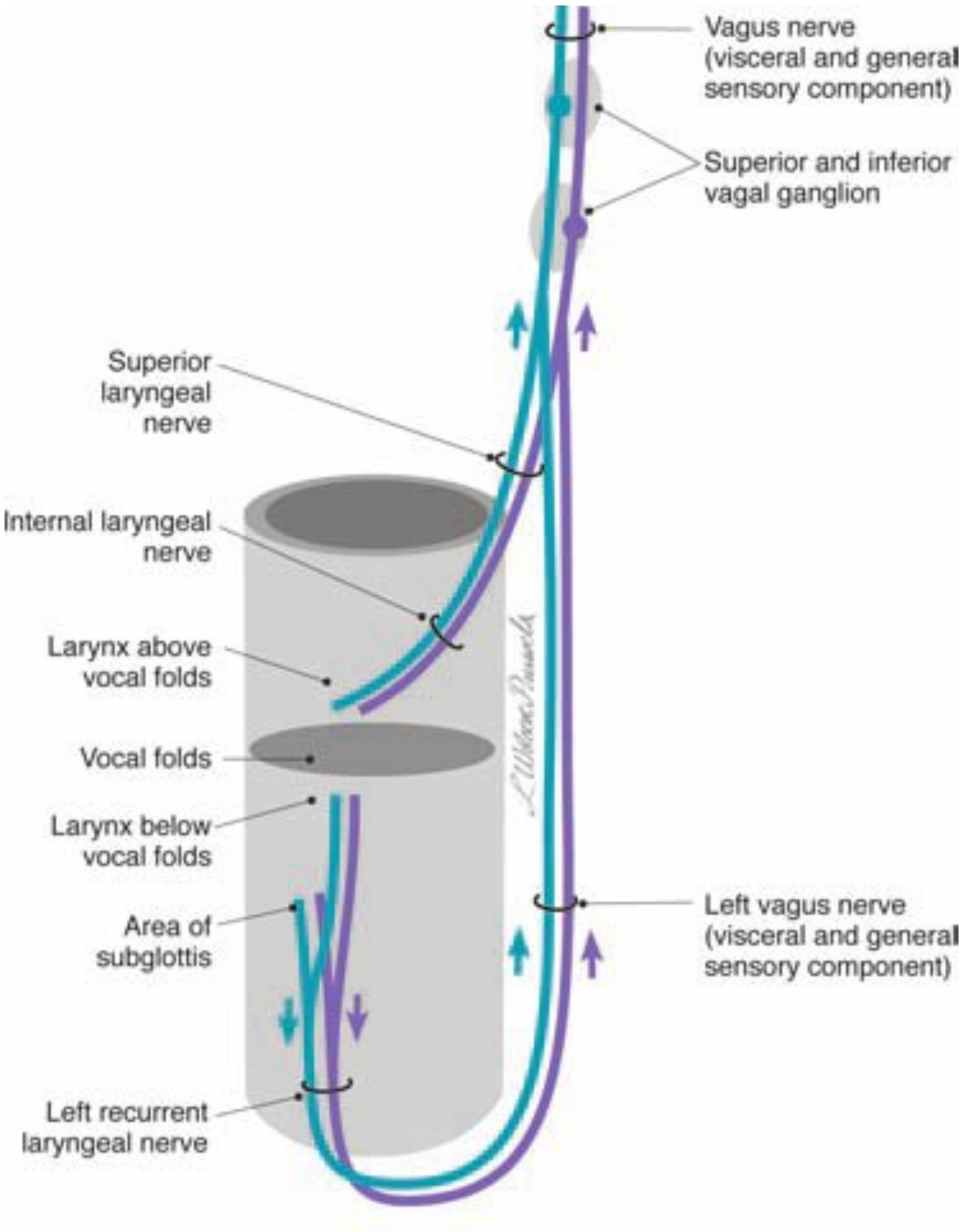
Vagus nerve (visceral and general sensory component)
Superior and inferior vagal ganglion
Superior laryngeal nerve
Internal laryngeal nerve
Larynx above vocal folds
Vocal folds
Larynx below vocal folds
Area of subglottis
Left recurrent laryngeal nerve
Left vagus nerve (visceral and general sensory component)
L. Wilson-Pauwels

FIGURE X–6 Schematic illustration depicting visceral and general sensory components of the left vagus nerve from the larynx.

The central processes pass upward through the jugular foramen and enter the medulla, then ascend in the spinal trigeminal tract to synapse in its nucleus (see Fig. X–5 inset). From the spinal trigeminal nucleus, second-order axons project via the medial lemniscus to the ventral posterior nucleus of the thalamus and on to the sensory cortex.

Second-order axons carrying pain signals enter the medulla and descend in the spinal trigeminal tract to end on the caudal part of the spinal trigeminal nucleus (see Fig. X–5 inset). From the nucleus, axons of secondary neurons cross the midline in the medulla and project to two distinct populations of neurons in the thalamus: neurons in the ventral medial posterior nucleus (VMpo) nucleus, which project to the postcentral sensory cortex (head region) for identification of the location and intensity of the pain; and neurons in the medial dorsal nucleus of the thalamus, which project to the cingulate cortex where the emotional component of pain is mediated.

VISCERAL SENSORY (AFFERENT) COMPONENT

Visceral sensation is carried in the visceral sensory component of the vagus nerve. It is not appreciated at a conscious level of awareness other than as "feeling

TABLE X–2 Branches of the Vagus Nerve (CN X)

Location	Branch	Modality			
		General sensory	Visceral sensory	Branchial motor	Parasympathetic motor
Jugular fossa	■ Meningeal	√			
	■ Auricular	√			
Neck	■ Pharyngeal	√	√	√	√
	■ Branches to aortic bodies		√		√
	■ Superior laryngeal	√	√	√	√
	Internal laryngeal	√	√		√
	External laryngeal			√	
	■ Recurrent laryngeal (right)	√	√	√	√
	■ Cardiac		√		√
Thorax	■ Cardiac		√		√
	■ Recurrent laryngeal (left)	√	√	√	√
	■ Pulmonary		√		√
	■ Esophageal		√		√
Abdomen	■ Gastrointestinal		√		√

good" or "feeling bad," unlike visceral pain that is carried in the sympathetic nervous system.

Visceral sensory fibers from plexuses around the abdominal viscera converge and join with the right and left gastric nerves of the vagus (Fig. X–7). These nerves pass upward through the esophageal hiatus (opening) of the diaphragm to merge with the plexus of nerves around the esophagus. Sensory fibers from plexuses around the heart and lungs also converge with the esophageal plexus and continue up through the thorax in the right and left vagus nerves.

The right and left vagus nerves are joined by nerves carrying visceral sensory information from the

- Baroreceptors (stretch receptors) in the aortic arch and chemoreceptors (monitoring pH, CO_2, and O_2 concentrations in the blood) in the aortic bodies (see Fig. X–7 inset).
- Larynx below the vocal cords in the recurrent laryngeal nerve (see Fig. X–6).
- Larynx above the vocal folds in the internal laryngeal nerve (see Fig. X–6).
- Mucous membrane of the epiglottis and base of the tongue in the pharyngeal plexus.

The central processes of the nerve cell bodies in the inferior vagal ganglion enter the medulla and descend in the tractus solitarius to enter the caudal part of the nucleus of the tractus solitarius. From the nucleus, bilateral connections important in the

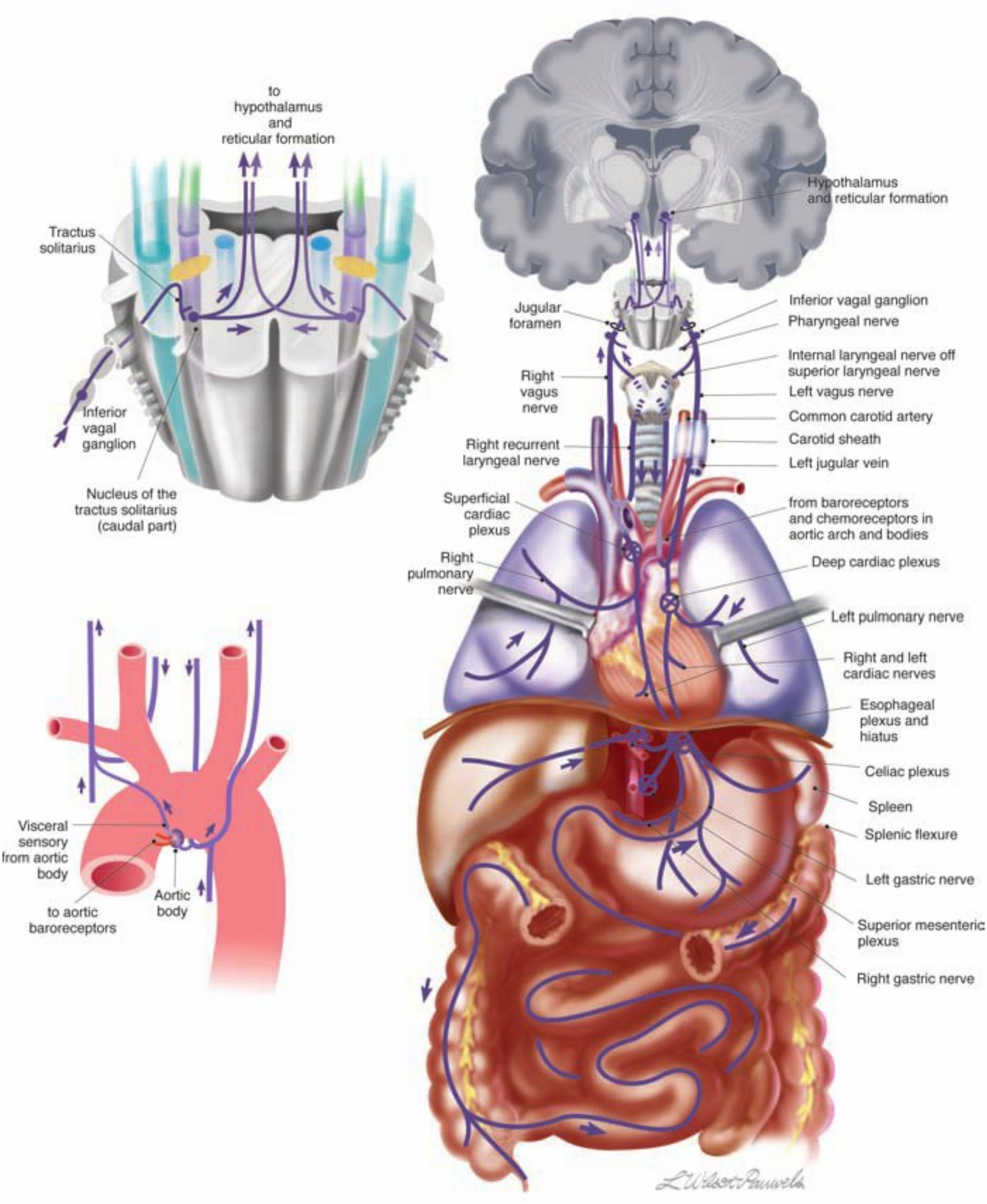
to hypothalamus and reticular formation
Tractus solitarius
Inferior vagal ganglion
Nucleus of the tractus solitarius (caudal part)
Visceral sensory from aortic body
to aortic baroreceptors
Aortic body
Hypothalamus and reticular formation
Jugular foramen
Inferior vagal ganglion
Pharyngeal nerve
Internal laryngeal nerve off superior laryngeal nerve
Right vagus nerve
Left vagus nerve
Common carotid artery
Right recurrent laryngeal nerve
Carotid sheath
Left jugular vein
Superficial cardiac plexus
from baroreceptors and chemoreceptors in aortic arch and bodies
Right pulmonary nerve
Deep cardiac plexus
Left pulmonary nerve
Right and left cardiac nerves
Esophageal plexus and hiatus
Celiac plexus
Spleen
Splenic flexure
Left gastric nerve
Superior mesenteric plexus
Right gastric nerve

FIGURE X–7 Visceral sensory component of the vagus nerve.

reflex control of cardiovascular, respiratory, and gastrointestinal functions are made with several areas of the reticular formation and the hypothalamus.

BRANCHIAL MOTOR (EFFERENT) COMPONENT

Bilateral corticobulbar fibers (fibers connecting the cortex with CN nuclei in the brain stem) are composed of axons from the premotor, motor, and other cortical areas. They descend through the internal capsule to synapse on motor neurons in the nucleus ambiguus, a column of cells just dorsal to the inferior olivary nucleus in the medulla (Fig. X–8). The nucleus ambiguus also receives sensory signals from other brain stem nuclei, mainly the spinal trigeminal and solitary nuclei, initiating reflex responses (e.g., coughing and vomiting). Lower motor neuron axons leave the nucleus ambiguus and travel laterally to leave the medulla as 8 to 10 rootlets. The caudal rootlets travel briefly with CN XI, rejoining with the rostral rootlets of CN X just below the inferior vagal ganglion (see Fig. X–3). The nerve leaves the skull through the jugular foramen to reach the constrictor muscles of the pharynx and the intrinsic muscles of the larynx (see Figs. X–1 and X–8).

The branchial motor fibers leave the vagus nerve in three major branches: pharyngeal, superior laryngeal, and recurrent laryngeal, all of which carry general and visceral sensation and parasympathetic visceral motor fibers as well.

Pharyngeal Branch

The pharyngeal branch, the principal motor nerve of the pharynx, traverses the inferior ganglion and passes inferomedially between the internal and the external carotid arteries. It enters the pharynx at the upper border of the middle constrictor and breaks up into the pharyngeal plexus to supply all the muscles of the pharynx and soft palate except the stylopharyngeus (CN IX) and tensor veli palatini (branchial motor component of CN V_3). Therefore, it supplies the superior, middle, and inferior constrictors, levator veli palatini, salpingopharyngeus, palatopharyngeus, and one muscle of the tongue, the palatoglossus (many are illustrated in Fig. X–8).

Superior Laryngeal Branch

The superior laryngeal nerve branches from the main trunk of the vagus nerve at the inferior vagal ganglion distal to the pharyngeal branch. It descends adjacent to the pharynx, dividing into internal (mainly sensory) and external (motor) laryngeal nerves. Branchial motor axons in the external laryngeal branch supply the cricothyroid muscles, and a few axons may supply part of the inferior constrictor.

It also sends branches to the pharyngeal plexus. The pharyngeal plexus, supplying the palate and pharynx, is formed by branches from the external laryngeal and pharyngeal nerves, as well as branches from CN IX and the sympathetic trunk (see Fig. X–8).

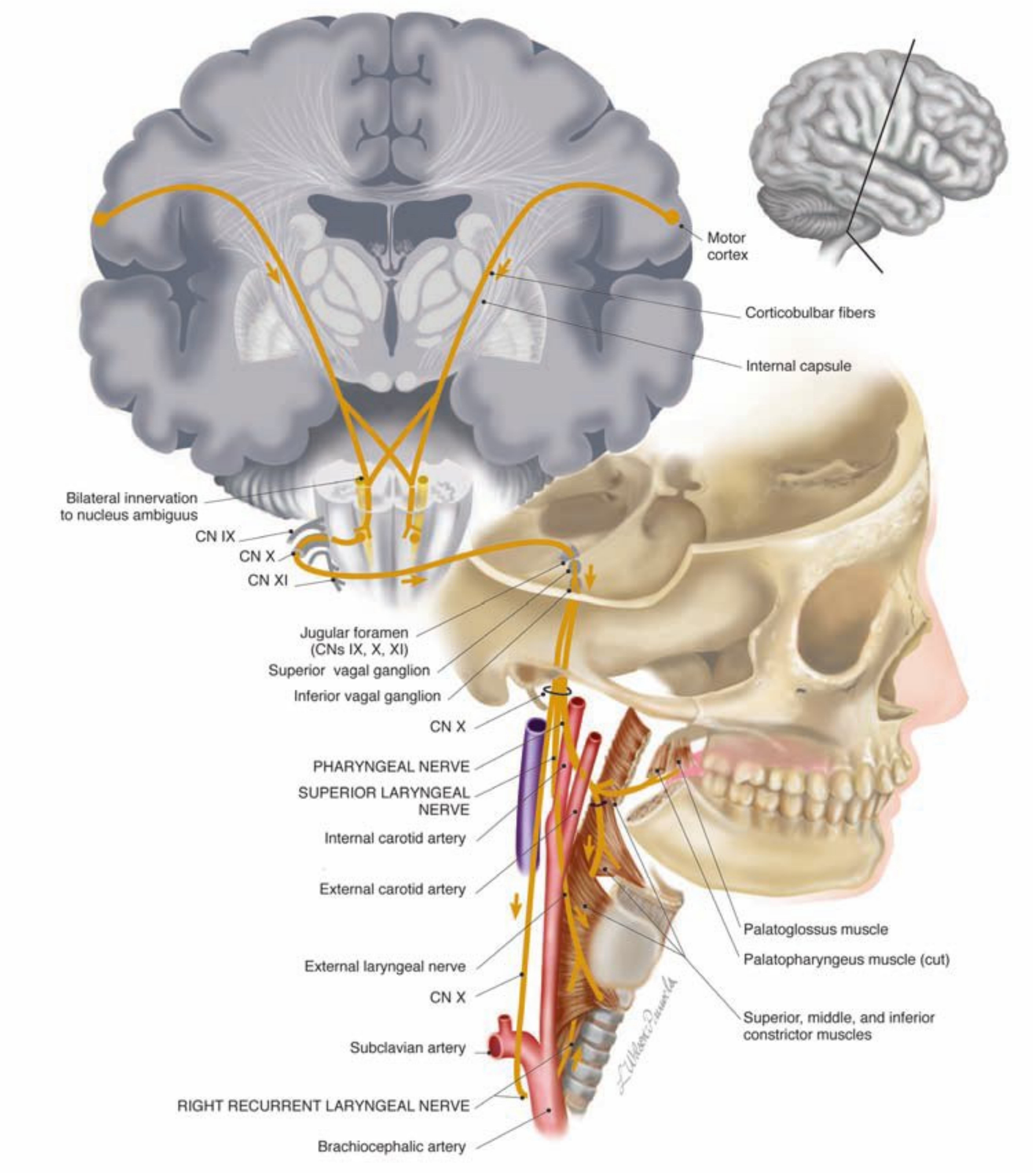

FIGURE X–8 Branchial motor component of the vagus nerve.

Recurrent Laryngeal Nerve

The recurrent laryngeal nerve, the third major branch, takes a different path on the right and left sides of the body (see Fig. X–4). The right recurrent laryngeal nerve arises from the vagus nerve anterior to the subclavian artery, then hooks back under the artery and ascends posterior to it in the groove between the trachea and the esophagus on the right side. The left recurrent laryngeal nerve arises from the left vagus on the aortic arch. It hooks back posteriorly under the arch and ascends through the superior mediastinum to reach the groove between the trachea and the esophagus on the left side. The recurrent nerves pass deep to the inferior margin of the inferior constrictor muscle. The branchial motor axons supply the intrinsic muscles of the larynx except the cricothyroid (see Fig. X–8).

PARASYMPATHETIC MOTOR (VISCERAL EFFERENT) COMPONENT

The parasympathetic nerve cell bodies of the vagus nerve are located in the dorsal motor nucleus of the vagus (Fig. X–9) and in the medial side of the nucleus

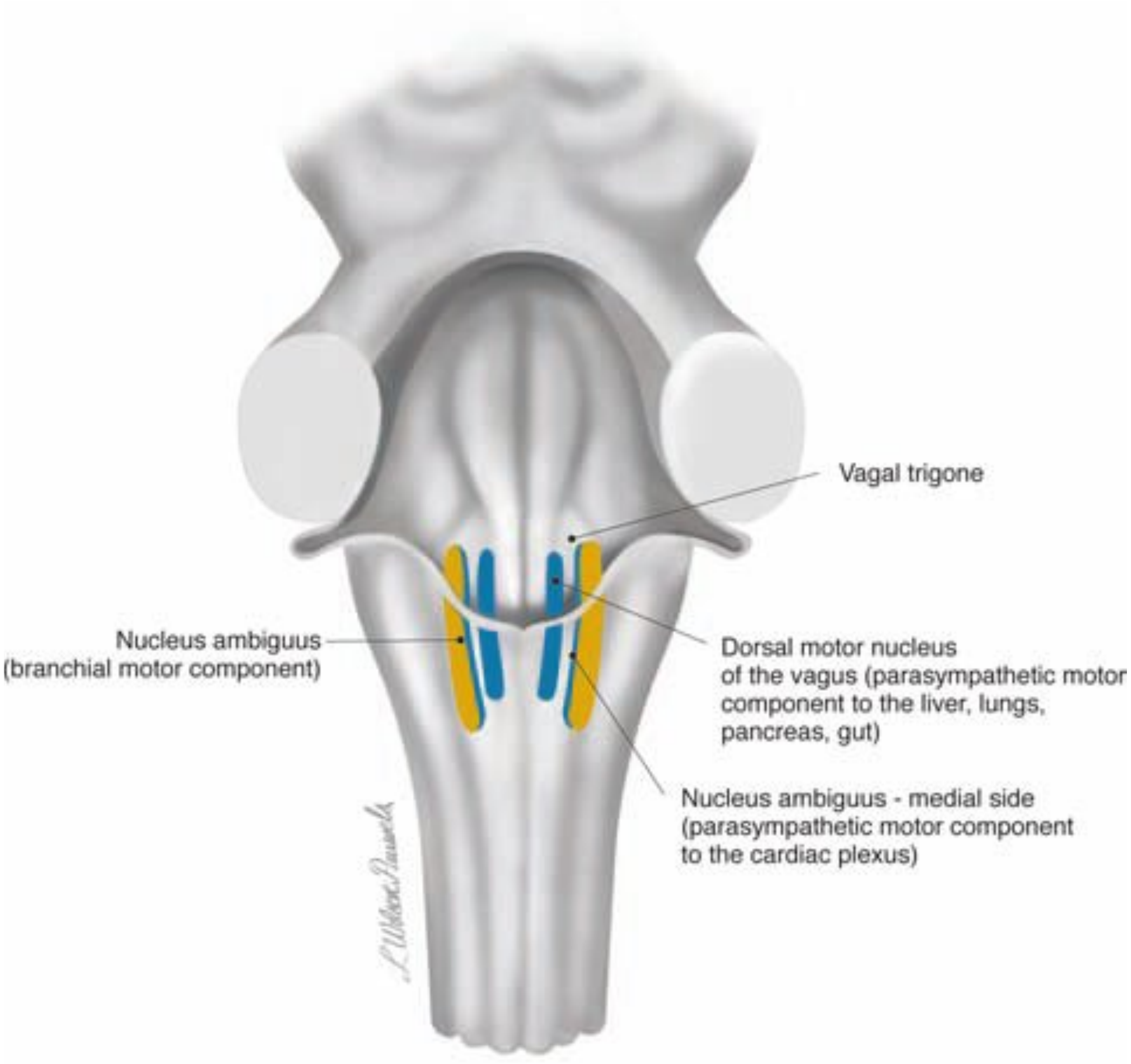
Vagal trigone
Nucleus ambiguus (branchial motor component)
Dorsal motor nucleus of the vagus (parasympathetic motor component to the liver, lungs, pancreas, gut)
Nucleus ambiguus - medial side (parasympathetic motor component to the cardiac plexus)
L Wilson-Pauwels

FIGURE X–9 Parasympathetic motor nuclei of the vagus nerve (posterior brain stem).

ambiguus. Neurons in the dorsal vagal nucleus innervate ganglia in the gut and its derivatives (lungs, liver, pancreas), whereas neurons in the nucleus ambiguus innervate ganglia in the cardiac plexus. They are all influenced by input from the hypothalamus, the olfactory system, the reticular formation, and the nucleus of the tractus solitarius. The dorsal motor nucleus of the vagus is located in the floor of the fourth ventricle (vagal trigone) and in the central gray matter of the closed medulla. Preganglionic fibers from this nucleus traverse the spinal trigeminal tract and nucleus and emerge from the lateral surface of the medulla to join the other components of the vagus nerve (see Figs. X–1 and X–2).

Within the pharynx and larynx, the vagal preganglionic axons activate ganglionic neurons that are secretomotor to the glands of the pharyngeal and laryngeal mucosa. Preganglionic axons are distributed to the pharyngeal plexus through the pharyngeal and internal laryngeal branches. Within the thorax, the vagi take different paths, but both break up into many branches that join plexuses around the major blood vessels to the lungs and the heart (Fig. X–10). Pulmonary branches cause bronchoconstriction, and esophageal branches act to speed up peristalsis in the esophagus by activating the smooth (nonstriated) muscle of the walls of the esophagus. The axons synapse in ganglia located in the walls of the individual organs. The cell bodies of cardiac preganglionic axons are located in the medial nucleus ambiguus. Their axons terminate on small ganglia associated with the heart and the aortic body in the arch of the aorta. They act to slow down the cardiac cycle.

The right and left gastric nerves emerge from the esophageal plexus. These nerves stimulate secretion by the gastric glands and are motor to the smooth muscle of the stomach. Intestinal branches act similarly on the small intestine, cecum, vermiform appendix, ascending colon, and most of the transverse colon. Within the walls of the gut, there are autonomic neurons that form a semi-independent network called the "enteric nervous system." This system can function reasonably well without input from sympathetic or parasympathetic divisions and is arranged in two distinct plexuses, the myenteric and submucosal plexuses of Auerbach and Meissner, respectively (Fig. X–11). The intestinal branches of the vagus synapse on ganglia within these plexuses and act to increase gut motility and secretion/absorption.

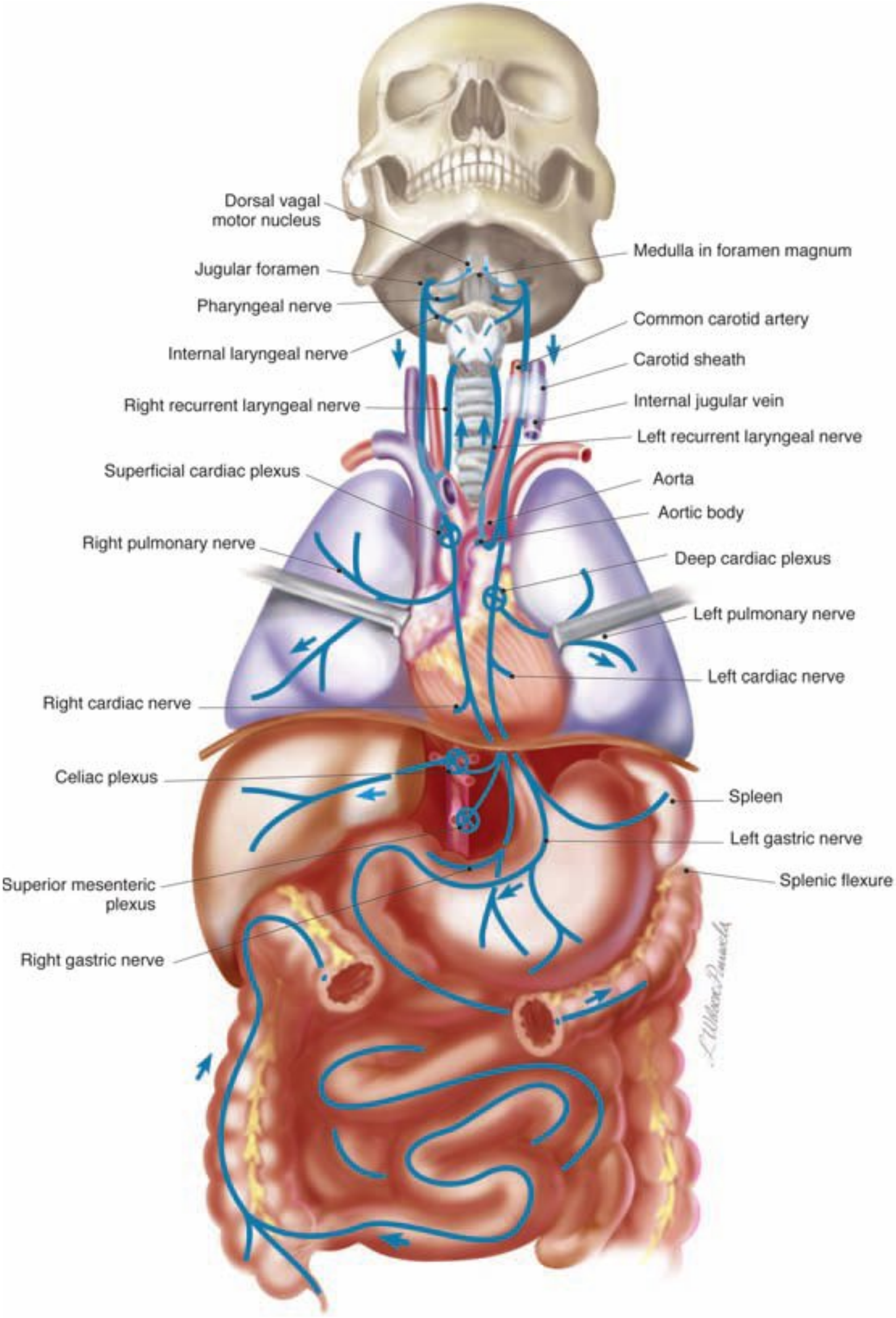

FIGURE X–10 Parasympathetic motor component of the vagus nerve.

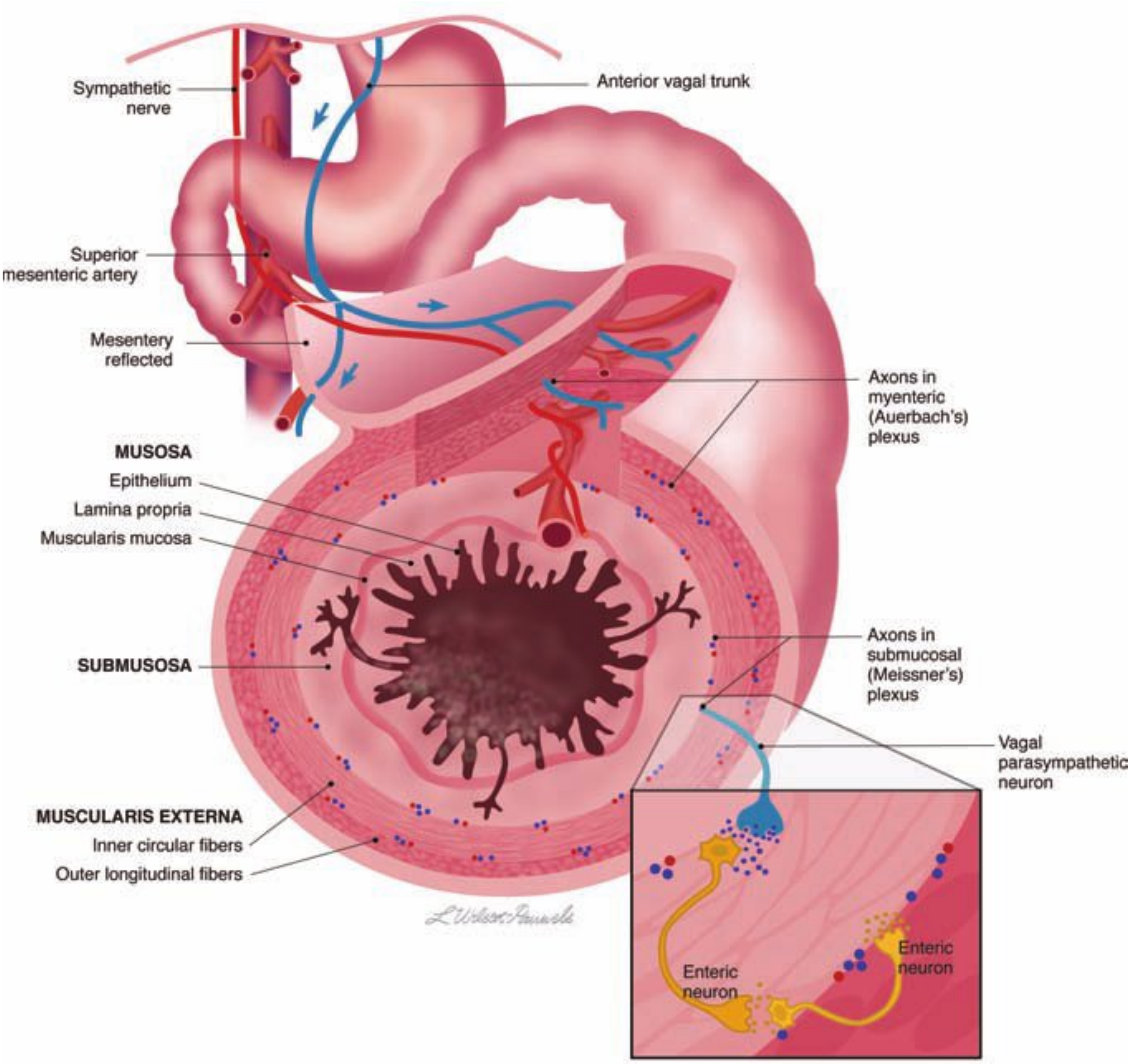

FIGURE X–11 The effect of the parasympathetic innervation of the vagus nerve on the enteric nervous system of the gut. The myenteric plexus (of Auerbach) between the inner circular and outer longitudinal muscle layers and the submucosal plexus (of Meissner) in the submucosal space are highlighted. The insert shows a simplified version of a parasympathetic nerve affecting an enteric neuron.

CASE HISTORY GUIDING QUESTIONS

1. What is a glomus jugulare tumor?
2. Why did Ruth hear a whooshing sound in her left ear?
3. Why did Ruth lose the gag reflex on her left side?
4. Why did Ruth develop a hoarse voice and have trouble swallowing?

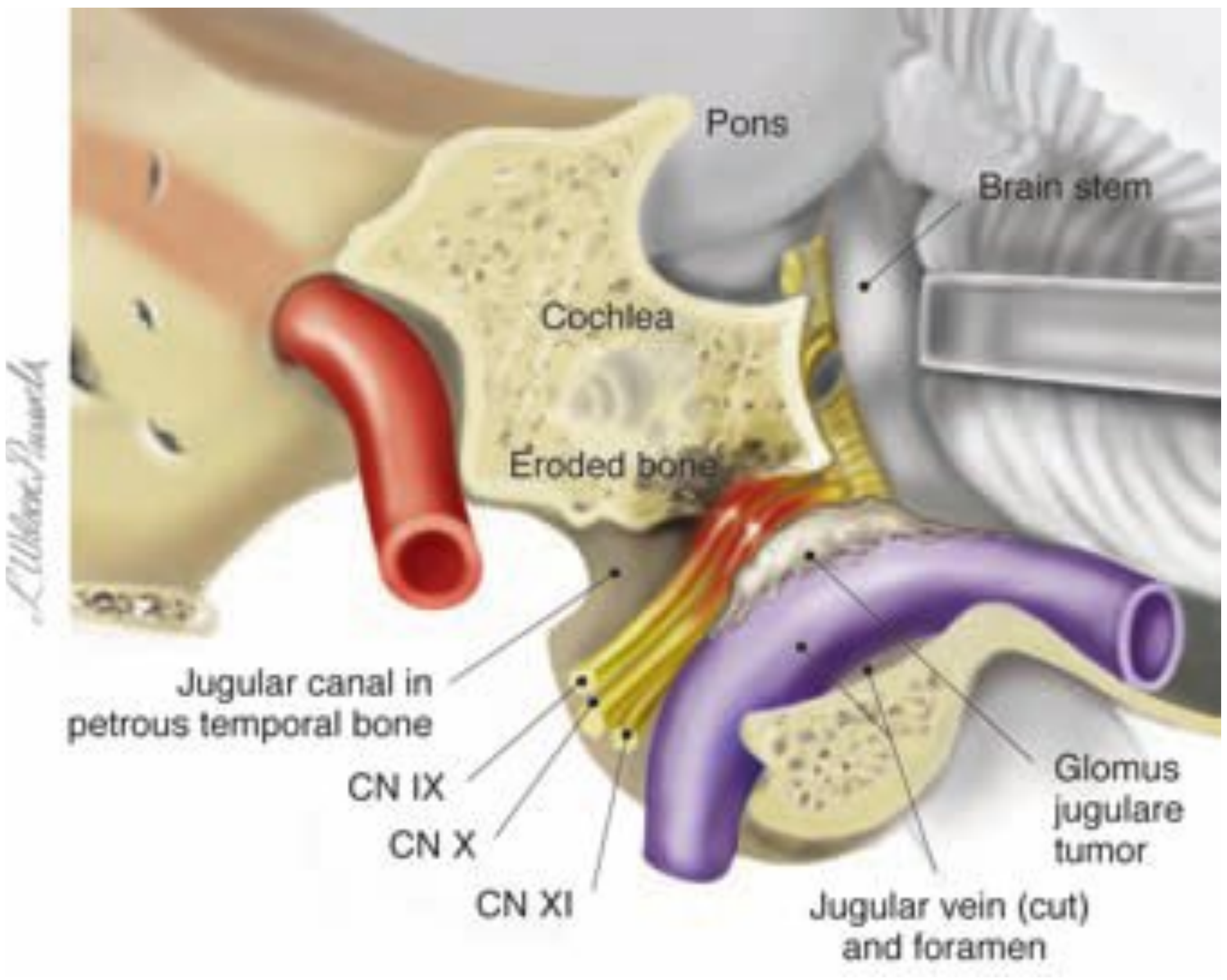
Pons
Brain stem
Cochlea
Eroded bone
Jugular canal in petrous temporal bone
CN IX
CN X
CN XI
Glomus jugulare tumor
Jugular vein (cut) and foramen

FIGURE X–12 Tumor of the glomus cells of the jugular bulb compressing cranial nerves IX, X, and XI (lateral view showing cut jugular foramen).

5. Why was Ruth's left sternomastoid muscle weakened?
6. What other clinical signs are seen in association with a glomus jugulare tumor?
7. Where along the course of CN X can a lesion occur?

1. What is a glomus jugulare tumor?

A glomus jugulare tumor is a tumor of the glomus cells of the jugular bulb, which is the proximal part of the internal jugular venous system (Fig. X–12). The glomus cells are paraganglia cells that are a part of the chemoreceptor system; therefore, like chemoreceptors found in the carotid bodies, they monitor O_2, CO_2, and pH. The tumor typically compresses CNs IX, X, and XI and erodes the jugular foramen. Women are affected more often than men, and the peak incidence is during middle adult life. The treatment involves surgical resection of the tumor and, in some cases, radiation therapy.

2. Why did Ruth hear a whooshing sound in her left ear?

This tumor is highly vascular and, therefore, has a robust blood flow. Because it is located immediately below the floor of the middle ear, sound from the turbulent blood flow passes through the bone and stimulates the cochlea, creating a perceived whooshing noise.

3. Why did Ruth lose the gag reflex on her left side?

The gag reflex involves the general sensory input from CN IX and the motor output of CN X (Figs. X–13, X–14 and IX–13). If either limb of the reflex arc is damaged, the gag reflex will be lost. In Ruth's case, the tumor compromised both the sensory (CN IX) and the motor (CNs V, X, and XII) limbs of the gag reflex.

4. Why did Ruth develop a hoarse voice and have trouble swallowing?

When the muscles controlling one of the vocal cords are paralyzed by loss of their innervation, the cord becomes lax and cannot vibrate against the other cord. As

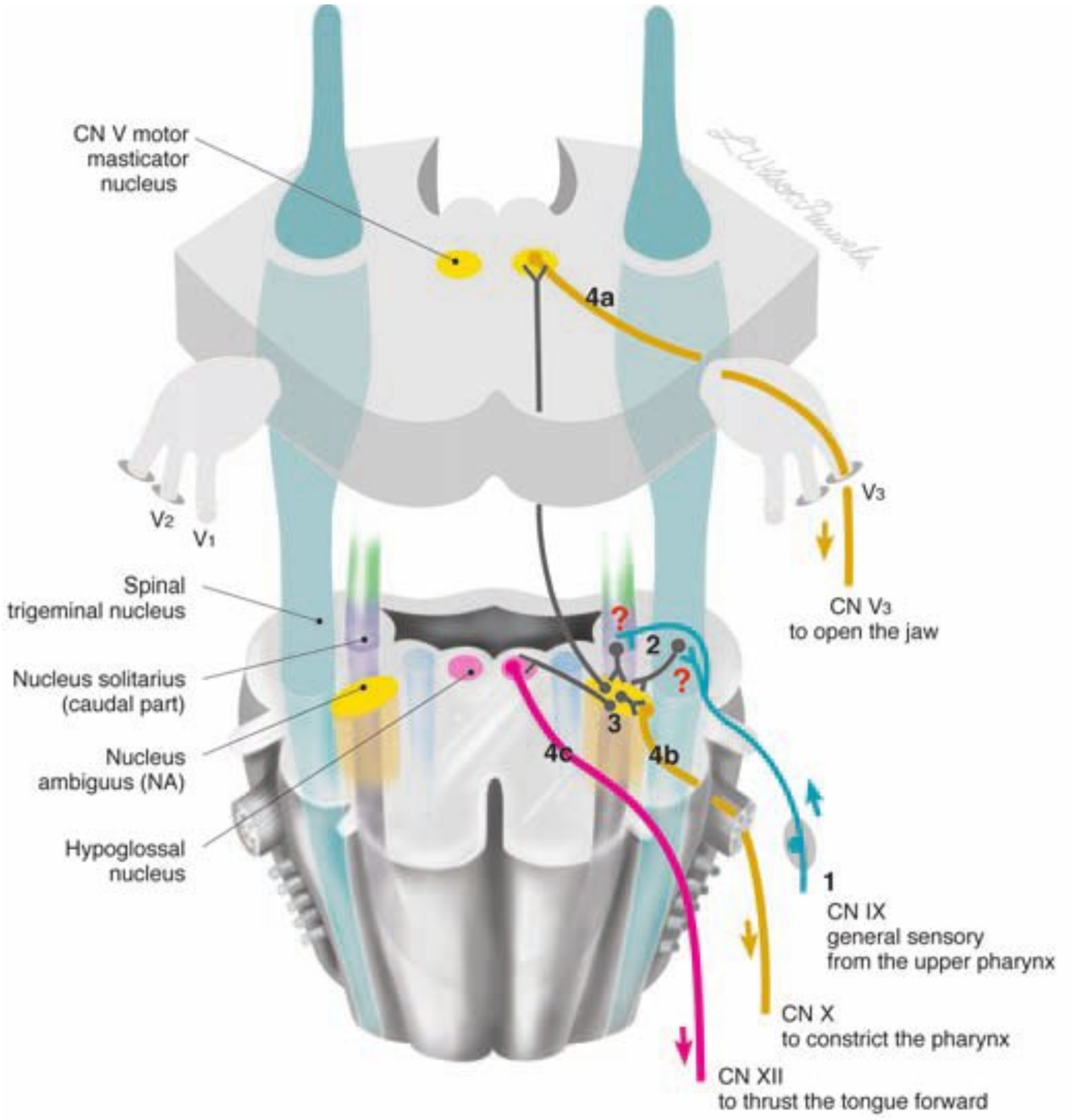

FIGURE X–13 Gag reflex pathways.

1. Stimulation of the upper pharynx activates CN IX sensory neurons.
2. Interneurons (grey) in the spinal trigeminal nucleus or in the caudal part of nucleus solitarius send signals to
3. Nucleus ambiguus that, in turn, activate:
4. a. neurons in the masticator nucleus that open the jaw via CN V_3,
   b. other neurons in nucleus ambiguus that cause pharyngeal constriction via CN X and
   c. neurons in the hypoglossal nucleus that cause thrusting of the tongue via CN XII.

NB. The pathways for the gag reflex in the human are not known with certainty. Since the pharynx is a point of transition between the body wall and the viscera, sensory afferents may be somatic, visceral or both. The afferent limb of the gag reflex, therefore, may synapse in the spinal trigeminal nucleus, the nucleus solitarius, or both (indicated with red question marks).

a result, the voice becomes low-pitched and hoarse. The patient, having to force increased amounts of air to set the intact cord in motion, becomes short of breath when speaking. Also, the impairment causes difficulty swallowing due to an inability to produce coordinated movements of the pharyngeal muscles. Difficulty in elevating the soft palate adequately (unilateral loss of levator palati muscle) would allow food to pass into the nasal cavity during swallowing.

5. Why was Ruth's left sternomastoid muscle weakened?

The sternomastoid muscle is innervated by the accessory nerve (CN XI) that exits the skull through the jugular foramen. The tumor has compressed and compromised the accessory nerve (see Fig. X–12).

6. What other clinical signs are seen in association with a glomus jugulare tumor?

The glomus jugulare tumor is an invasive tumor that has multiple extensions and will spread into any opening or fissure in the petrous temporal bone. Clinical signs are the result of invasion of the tumor and compression of adjacent nerves. The typical syndrome consists of a systolic bruit (abnormal sound or murmur), dysphagia (difficulty in swallowing), and dysphonia (difficulty in speaking) because of damage to CNs VII, IX, and X.

Other neurologic findings are correlated with the extension of the tumor. If the tumor

- Spreads toward the foramen magnum, there may be a CN XII palsy (paresis or paralysis of tongue muscles).

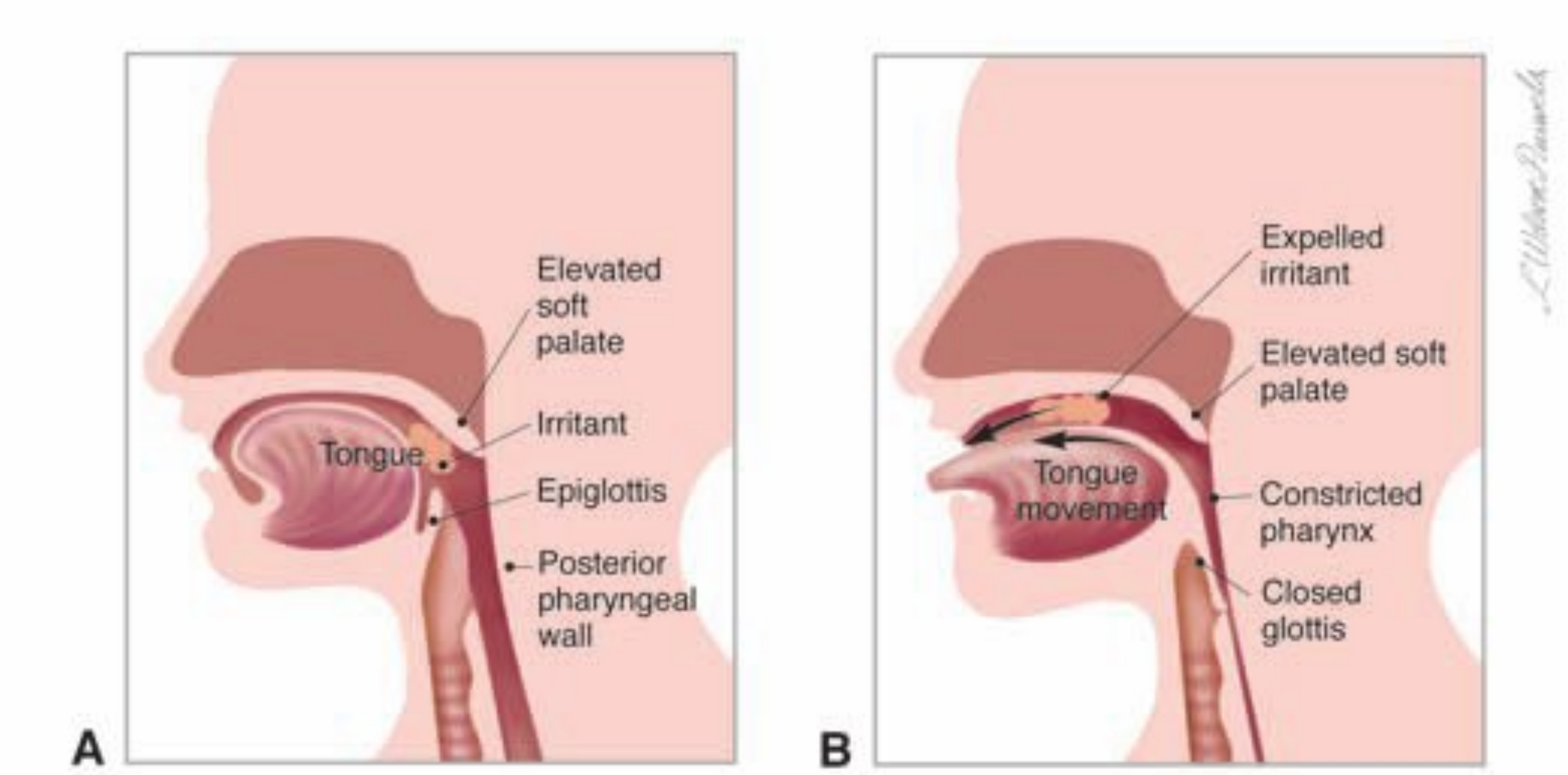

FIGURE X–14 The gag reflex. **A**, Irritant in the oropharynx stimulates the posterior tongue. As a result, cranial nerve IX's general sensory afferent nerve fibers are stimulated. **B**, A reflex response by cranial nerve X cell bodies in the nucleus ambiguus stimulates branchial motor efferent nerves resulting in elevation of the soft palate, closure of the glottis by the epiglottis, and contraction of the pharyngeal wall to expel the foreign object.

- Invades the intrapetrous carotid canal, a Horner's syndrome, characterized by miosis (constricted pupil), ptosis (drooping of the eyelid), enophthalmos (recession of the eyeball), and redness and dry skin on the ipsilateral face, may develop due to sympathetic nerve involvement.
- Spreads into the inner ear, there may be vestibular (balance) and cochlear (diminished hearing) involvement.
- Extends directly into the external auditory canal, a vascular tumor may be visualized.

7. Where along the course of CN X can a lesion occur?

A lesion of the vagus nerve can occur anywhere along its course from the cortex to the organ of innervation. An upper motor neuron lesion (UMNL) can occur anywhere between the cortex and the nucleus ambiguus. Lesions that involve these fibers are typically ischemias (insufficient blood supply), infarcts, or tumors. Bilateral UMNLs involving the corticobulbar tracts affect the bulbar[‡] musculature and are referred to as a "pseudobulbar palsy." This is a misleading term because there is nothing "pseudo" about a palsy. "Spastic bulbar palsy" would be a better term.

A lesion at the level of the nucleus ambiguus and below is a lower motor neuron lesion (LMNL). A unilateral lesion of the lower motor neuron fibers results in a bulbar palsy (light or incomplete paralysis) of the bulbar muscles on the ipsilateral side. LMNLs can occur from mass lesions compressing the pons, from tumors of the jugular foramen, and from surgical mishaps following procedures in the neck area such as a carotid endarterectomy or a thyroidectomy. The LMNLs can occur also from compression of the left recurrent laryngeal nerve by lung tumors or paratracheal lymph nodes compressing the nerve as it passes through the thorax.

CLINICAL TESTING

CN X is usually tested in conjunction with CN IX by assessment of the gag reflex. However, it is possible to test CN X in isolation. A unilateral lesion of the nerve results in lowering and flattening of the palatal arch on the affected side. Therefore, when examining CN X, one should observe the posterior pharynx at rest and then on phonation. On phonation (say "Ah."), there is contraction of the superior pharyngeal constrictor muscle. Unilateral paresis of the superior pharyngeal constrictor results in deviation of the uvula to the normal side and also a pull of the posterior pharyngeal wall toward the intact side (see Fig. X–15). Because the damaged side

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[‡] The term "bulbar" means a swelling. In neurology, we use the term "bulbar" to refer to the medulla and/or brain stem. Because corticobulbar tracts are those that go from the cortex to the brain stem to synapse on nuclei there, they are named "corticobulbar." Muscles supplied by these nerves are called "bulbar" muscles.

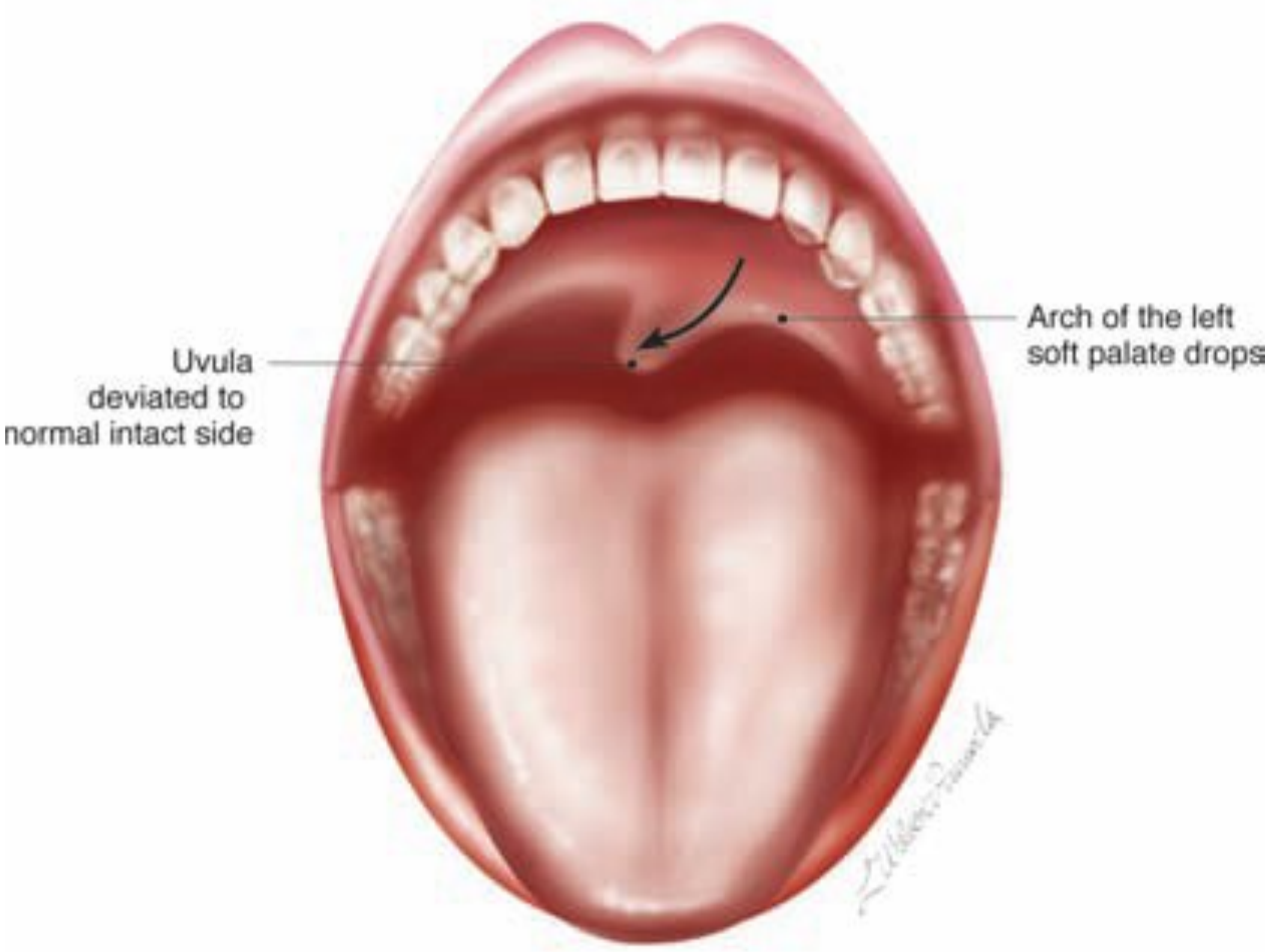

FIGURE X–15 Lower motor neuron lesion (LMNL) on the patient's left side.

cannot counter the pull of the intact side, the motion resembles that of swagging or drawing a curtain (of pharyngeal muscles) to the unaffected side, as you would swag a drape to the side. (See also "Cranial Nerves Examination" on CD-ROM.)

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XI

Accessory Nerve

CASE HISTORY

Burt, a 55-year-old businessman, was having episodes of weakness involving his right face and arm. These were diagnosed as transient ischemic attacks (TIAs), often referred to as "reversible strokes." During investigation for his TIAs, he was found to have 90% narrowing of his left internal carotid artery and underwent left carotid endarterectomy.

Approximately 2 weeks postoperative, Burt began to notice he was having problems pulling a sweater off over his head and was unable to bring his left arm over his head while swimming. He also developed a constant aching on the left side of his neck and left ear and a dull pain in his left shoulder. The vascular surgeon referred Burt to a neurologist. The neurologist noted that sensation was intact over Burt's face, neck, and shoulders, but he had weakness with shoulder elevation on his left side and was unable to abduct (raise) his left arm above the level of his shoulder. Electromyographic and nerve conduction studies showed that there had been damage to the branch of the left accessory nerve that supplies the upper fibers of the trapezius muscle but that the branch to the sternomastoid* muscle had been spared.

ANATOMY OF THE ACCESSORY NERVE

Information from the premotor association cortex and other cortical areas is fed into the motor cortex by association fibers. Axons of cortical neurons descend in the corticospinal[†] tract through the posterior limb of the internal capsule. Cortical neurons destined to supply the sternomastoid muscle descend to the ipsilateral

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*"Sternomastoid" is a shortened form of "sternocleidomastoid: and is used in this text.

[†] The term "corticospinal tract" versus "corticobulbar tract" is used because the descending axons project to the accessory nucleus in the spinal cord rather than to a nucleus in the brain stem.

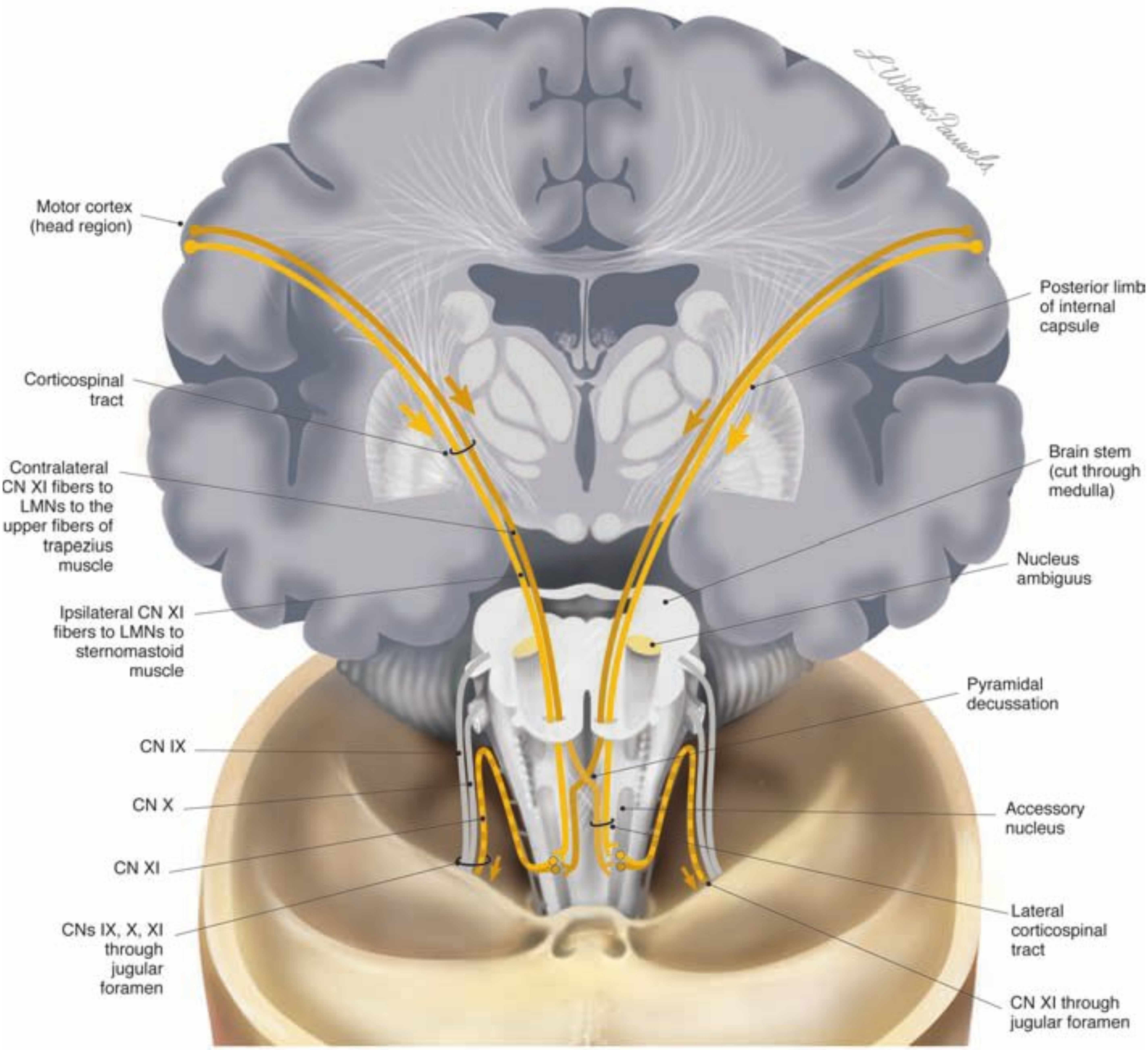
L Wilson-Pauwels
Motor cortex (head region)
Corticospinal tract
Contralateral CN XI fibers to LMNs to the upper fibers of trapezius muscle
Ipsilateral CN XI fibers to LMNs to sternomastoid muscle
CN IX
CN X
CN XI
CNs IX, X, XI through jugular foramen
Posterior limb of internal capsule
Brain stem (cut through medulla)
Nucleus ambiguus
Pyramidal decussation
Accessory nucleus
Lateral corticospinal tract
CN XI through jugular foramen

FIGURE XI–1 Branchial motor component of cranial nerve XI demonstrating ipsilateral innervation to LMNs innervating the sternomastoid muscle (yellow) and contralateral innervation to LMNs innervating the upper fibers of the trapezius muscle (ochre). The brain stem is elevated. LMNs = lower motor neurons.

accessory nucleus located in the lateral part of the anterior gray column of the upper five or six segments of the cervical spinal cord (Fig. XI–1), approximately in line with the nucleus ambiguus. Axons designated to supply the upper fibers of the trapezius muscle cross the midline in the pyramidal decussation to synapse in the contralateral accessory nucleus (Table XI–1; see also Fig. XI–1).

Various authors have described the supranuclear input to the accessory nucleus as entirely ipsilateral to the sternomastoid muscle and contralateral to the upper fibers

TABLE XI–1 Component, Nucleus, and Function of the Accessory Nerve (CN XI)

Component	Nucleus	Function
Branchial motor (efferent)	Accessory nucleus	To innervate sternomastoid and upper fibers of trapezius muscles

of trapezius, whereas others have described the input to sternomastoid as primarily ipsilateral with the possibility of a double crossing of the fibers from the hemisphere to the accessory nucleus (*Gray's Anatomy*, 2008; Brazis et al., 2006). This may explain why, in surgical neck dissections for cancer, patients have varied results.

From the accessory nucleus, postsynaptic fibers emerge from the lateral white matter of the spinal cord as a series of rootlets to form the accessory nerve (Fig. XI–2 and Chapter X, Fig. X–2). The rootlets emerge posterior to the ligamentum denticulatum but anterior to the dorsal roots of the spinal cord (see Fig. XI–2). The rootlets form a nerve trunk that ascends rostrally in the subarachnoid space and parallel to the spinal cord as far as the foramen magnum. At the foramen magnum,

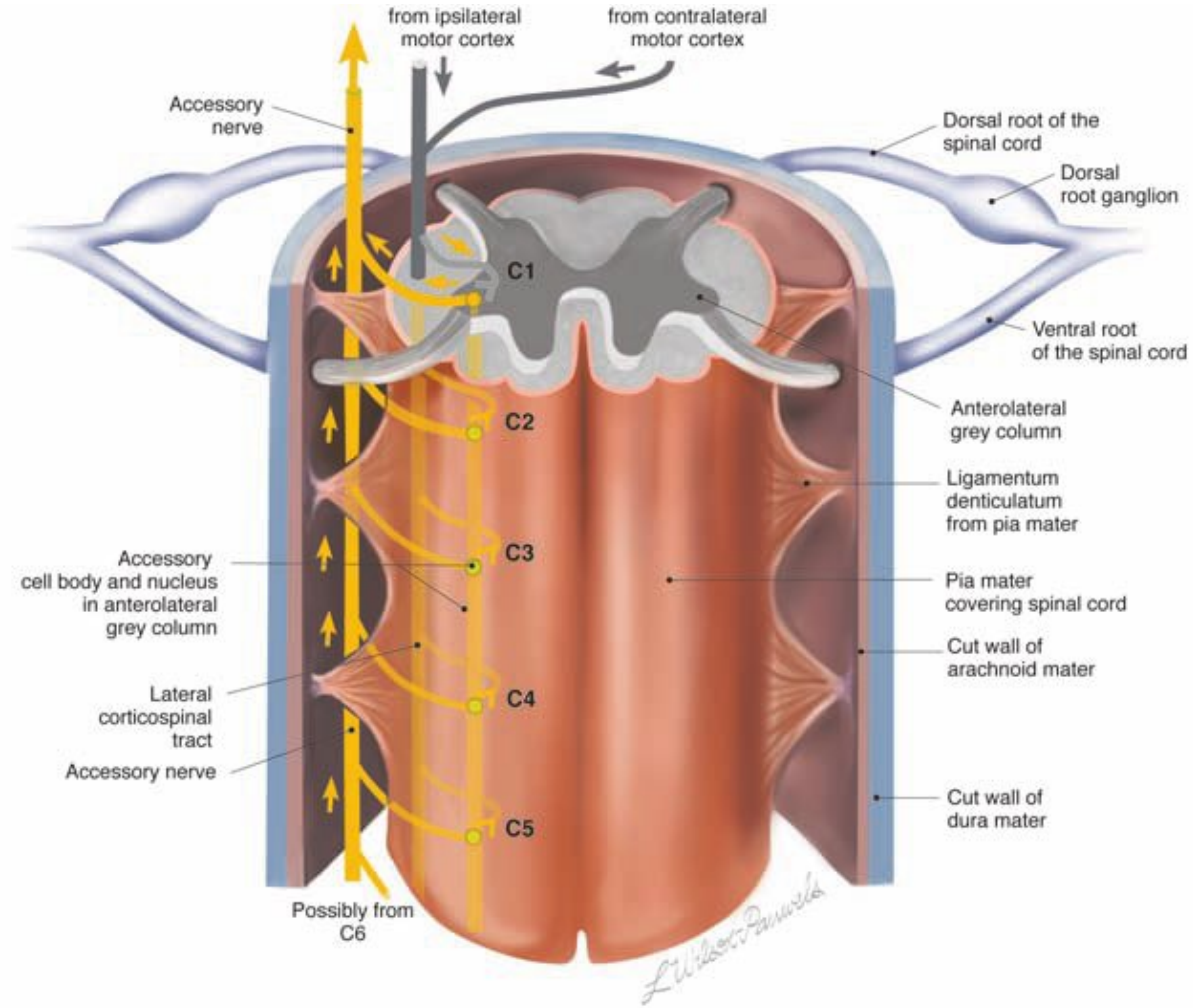
from ipsilateral motor cortex
from contralateral motor cortex
Accessory nerve
Dorsal root of the spinal cord
Dorsal root ganglion
C1
Ventral root of the spinal cord
C2
Anterolateral grey column
Ligamentum denticulatum from pia mater
C3
Accessory cell body and nucleus in anterolateral grey column
Pia mater covering spinal cord
Cut wall of arachnoid mater
C4
Lateral corticospinal tract
Accessory nerve
C5
Cut wall of dura mater
Possibly from C6

FIGURE XI–2 Branchial motor component of the accessory nerve—C1 to C5 or C6.

the nerve passes posterior to the vertebral artery to enter the posterior cranial fossa. The fibers join with caudal fibers from cranial nerve (CN) X and then separate from them within the jugular foramen (see Chapter X, Fig. X–2).

As the accessory nerve emerges from the jugular foramen, it passes posteriorly, medial to the styloid process, descends obliquely, and enters the upper portion of the sternomastoid muscle on its deep surface. Some of the fibers terminate in this muscle, and the remaining fibers pass through the muscle to emerge at the midpoint of its posterior border. These fibers then cross the posterior triangle of the neck, superficial to the levator scapulae, where they are closely related to the superficial cervical lymph nodes. Five centimeters above the clavicle, the nerve passes deep to the anterior border of the trapezius to supply the upper fibers of this muscle (Fig. XI–3).

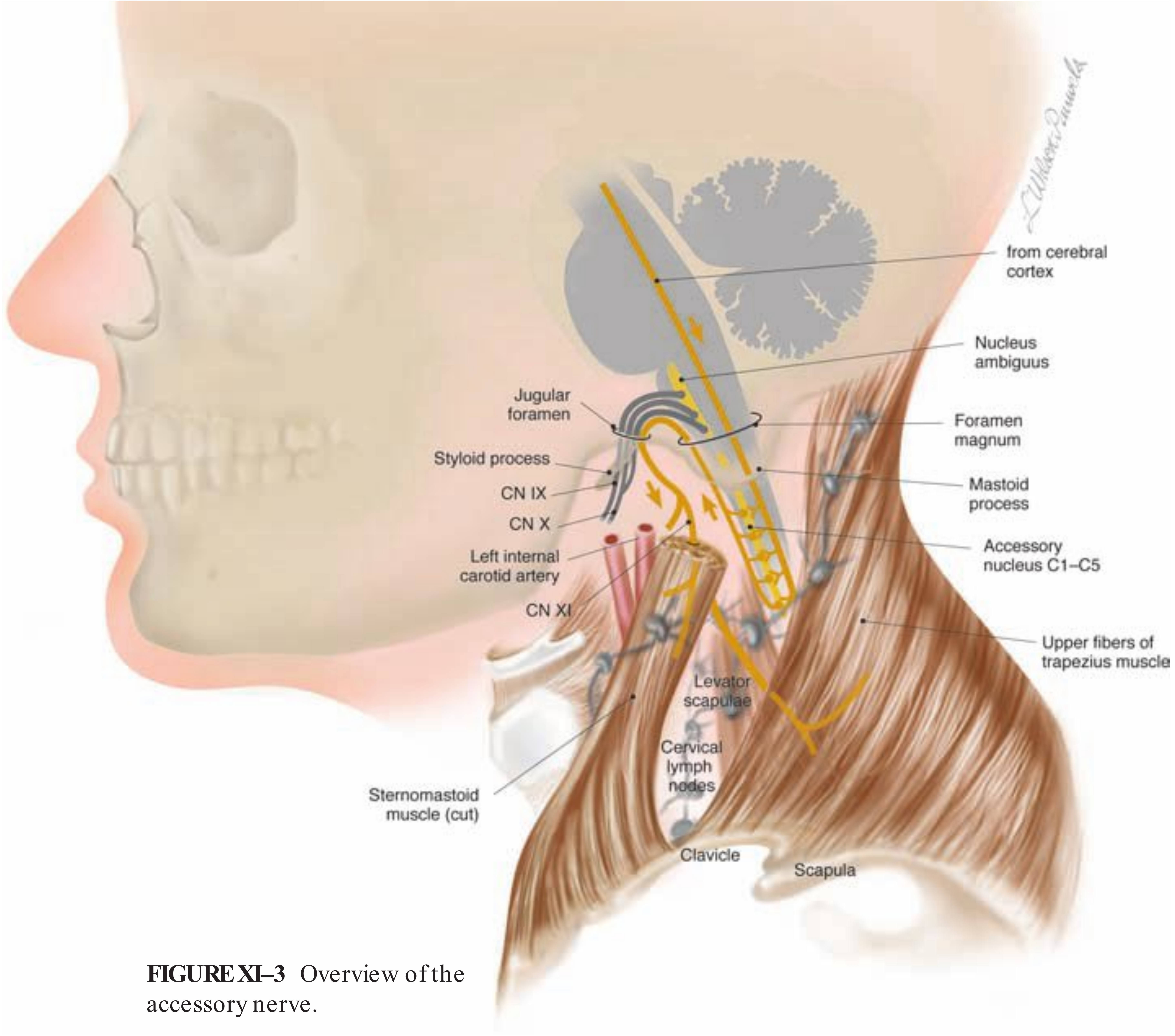

FIGURE XI–3 Overview of the accessory nerve.

Accessory Nucleus

The accessory nucleus is considered by some authors to be somatic motor to the sternomastoid and trapezius muscles. Others describe it as branchial motor to these same muscles, and still others describe it as branchial motor to sternomastoid and somatic motor to trapezius. In this book, the nucleus is considered to be branchial motor because it occupies a position in the ventral horn that is in line with other branchial motor nuclei. In addition, its rootlets exit from the cord in the same position as other branchial motor rootlets (i.e., between somatic motor and sensory rootlets).

Accessory Nerve

In this text, the "accessory nerve" is defined as the axons of only those lower motor neurons (LMNs) that emerge from the accessory nucleus (C1–5). Some other textbooks describe the accessory nerve as having both a rostral/cranial root (axons from the nucleus ambiguus traveling with CN X) and a caudal/spinal root (axons from the accessory nucleus, CN XI). See Chapter X and Fig. X–2 for a further explanation.

CASE HISTORY GUIDING QUESTIONS

1. What is the function of the sternomastoid muscle?
2. Why are the right sternomastoid and left trapezius muscles controlled by the same side of the cortex?
3. In Burt's case, how did his CN XI get damaged? Why is his trapezius muscle affected and his sternomastoid muscle spared?
4. What else could cause an isolated accessory nerve palsy?
5. If the accessory nerve is strictly motor in function, why is Burt experiencing pain?

1. What is the function of the sternomastoid muscle?

The sternomastoid muscle's upper attachment is to the mastoid process and the lateral half of the superior nuchal line of the skull. Its lower attachment is to the medial one-third of the clavicle and by a separate head to the manubrium of the sternum (Fig. XI–4). Therefore, the muscle acts to pull the mastoid process toward the clavicle, resulting in a rotation of the head and an elevation of the chin to the opposite side (see Fig. XI–4). The LMNs that innervate the right sternomastoid muscle receive ipsilateral input from the right cortex via the accessory nerve (CN XI) (see Fig. XI–1).

2. Why are the right sternomastoid and left trapezius muscles controlled by the same side of the cortex?

When the left upper limb muscles (including trapezius) are being used to manipulate an object, the head is turned to the left side to see what is happening. It is the

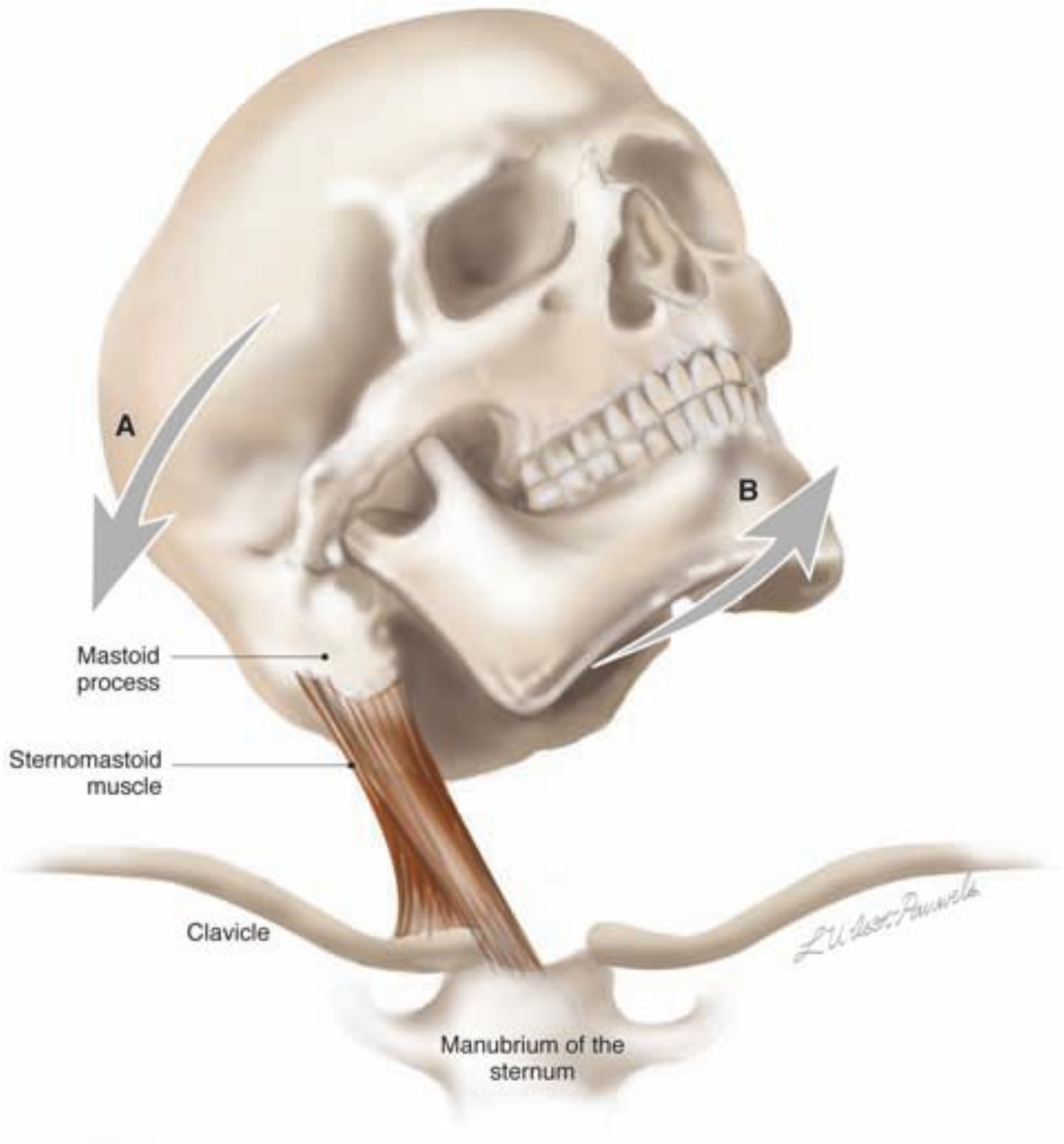

FIGURE XI–4 Action of the right sternomastoid muscle. **A**, the muscle pulls the mastoid process toward the clavicle resulting in **B**, the head rotating and the chin elevating to the opposite side.

Right cortex ⟶ Ipsilateral LMNs to right sternomastoid muscle ⟶ Chin elevates and head turns to left

Right cortex ⟶ Contralateral LMNs to left upper trapezius muscle ⟶ Left shoulder elevates

right sternomastoid muscle that turns the head to the left. However, it is the left upper trapezius muscle that elevates the shoulder. Therefore, the right cortex controls all the muscles necessary to perform these actions.

3. In Burt's case, how did his CN XI get damaged? Why is his trapezius muscle affected and his sternomastoid muscle spared?

The accessory nerve usually gives off branches to the sternomastoid prior to entering the muscle. The nerve then traverses the muscle and crosses the posterior triangle of the neck to reach and supply the upper fibers of trapezius muscle (see Fig. XI–3).

During Burt's carotid endarterectomy surgery, the sternomastoid muscle was retracted to fully expose the common carotid artery. If the retraction is too vigorous, it has the potential to stretch and selectively damage the branch of the accessory nerve to the trapezius lying within the sternomastoid muscle. This is what happened to Burt.

4. What else could cause an isolated accessory nerve palsy?

An isolated accessory nerve palsy is uncommon; however, the accessory nerve can be damaged during surgical procedures on the neck (e.g., lymph node biopsy, cervicofacial lift, or internal jugular vein cannulation). In addition, trauma, including carrying heavy loads on the shoulder, attempted suicide by hanging, and even neck bites sustained during passionate sex, can damage the accessory nerve. It is the axons that supply the trapezius muscle that are the most vulnerable to damage, because they take a long course through the posterior triangle of the neck (see Fig. XI–3). The axons to the sternomastoid muscle, which leave the main trunk high in the neck, are relatively spared.

5. If the accessory nerve is strictly motor in function, why is Burt experiencing pain?

Burt is experiencing pain because his inactive trapezius muscle is no longer supporting his shoulder, and the remaining shoulder and arm muscles are required to assume an unaccustomed load. The resulting muscle fatigue and strain on the muscles and ligaments results in pain in his left shoulder, lateral neck, and periauricular regions. His pain can be relieved by support of the arm at the elbow with a sling.

CLINICAL TESTING

The accessory nerve is a purely motor nerve that supplies the sternomastoid and trapezius muscles. Each of these muscles should be tested separately because, as in Burt's case, it is possible to have a partial accessory nerve palsy affecting only one of these muscles.

The initial step involved in testing the accessory nerve is the assessment of the bulk of the sternomastoid and trapezius muscles. If there has been damage to the nerve (LMN lesion) there may be evidence of wasting of the muscle. Once muscle bulk is assessed, strength should be tested.

Assessment of the Sternomastoid Muscle

The sternomastoid muscle functions to tilt the face and elevate the chin to the opposite side. Therefore, by asking the patient to tilt the face up and toward the opposite side, one can assess the bulk of the muscle by viewing and palpating it

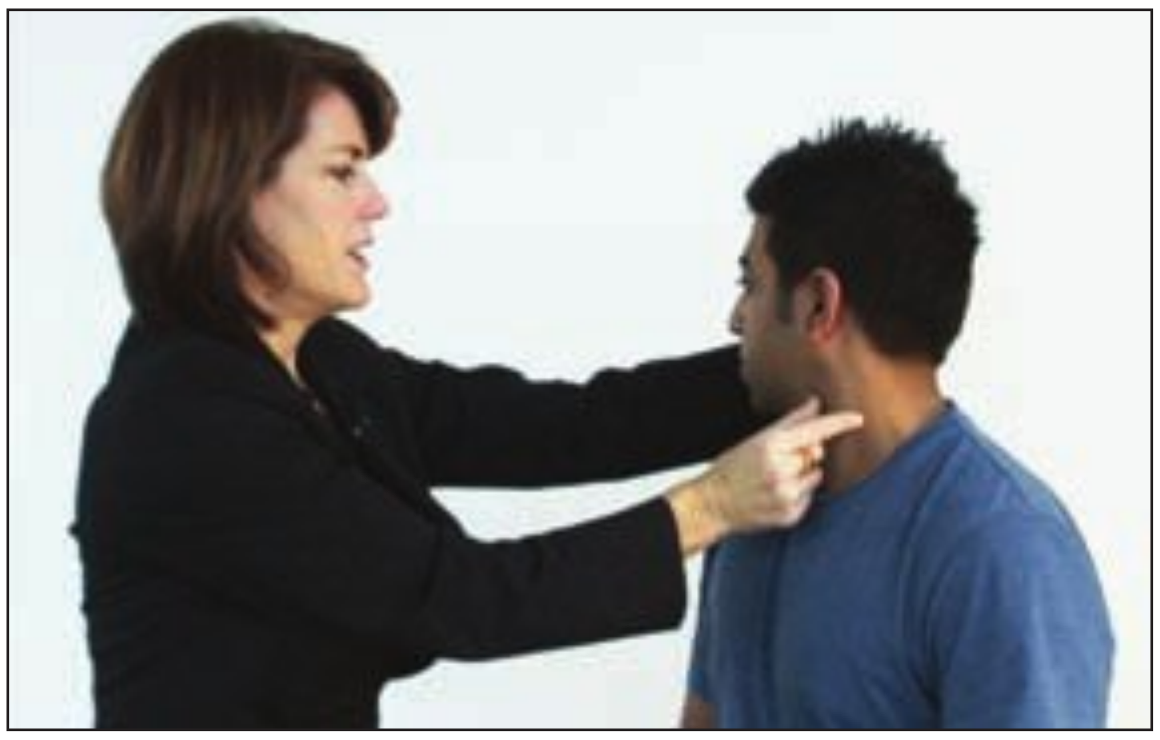

FIGURE XI–5 Examination and palpation of the left sternomastoid muscle.

(Fig. XI–5). The strength of this muscle is assessed by asking the patient to tilt the chin toward the opposite side against resistance.

Assessment of the Trapezius Muscle

The trapezius muscle functions to retract the head and to elevate, retract, and rotate the scapula. To assess the bulk of this muscle, it is important to expose the shoulder fully. A wasted trapezius muscle may result in a downward and lateral rotation of the scapula and some shoulder drop on the affected side. This can be seen when looking at the patient's back (Fig. XI–6).

The innervation of the trapezius muscle is variably described, but it is generally agreed that the upper fibers of the trapezius are innervated by the accessory nerve.

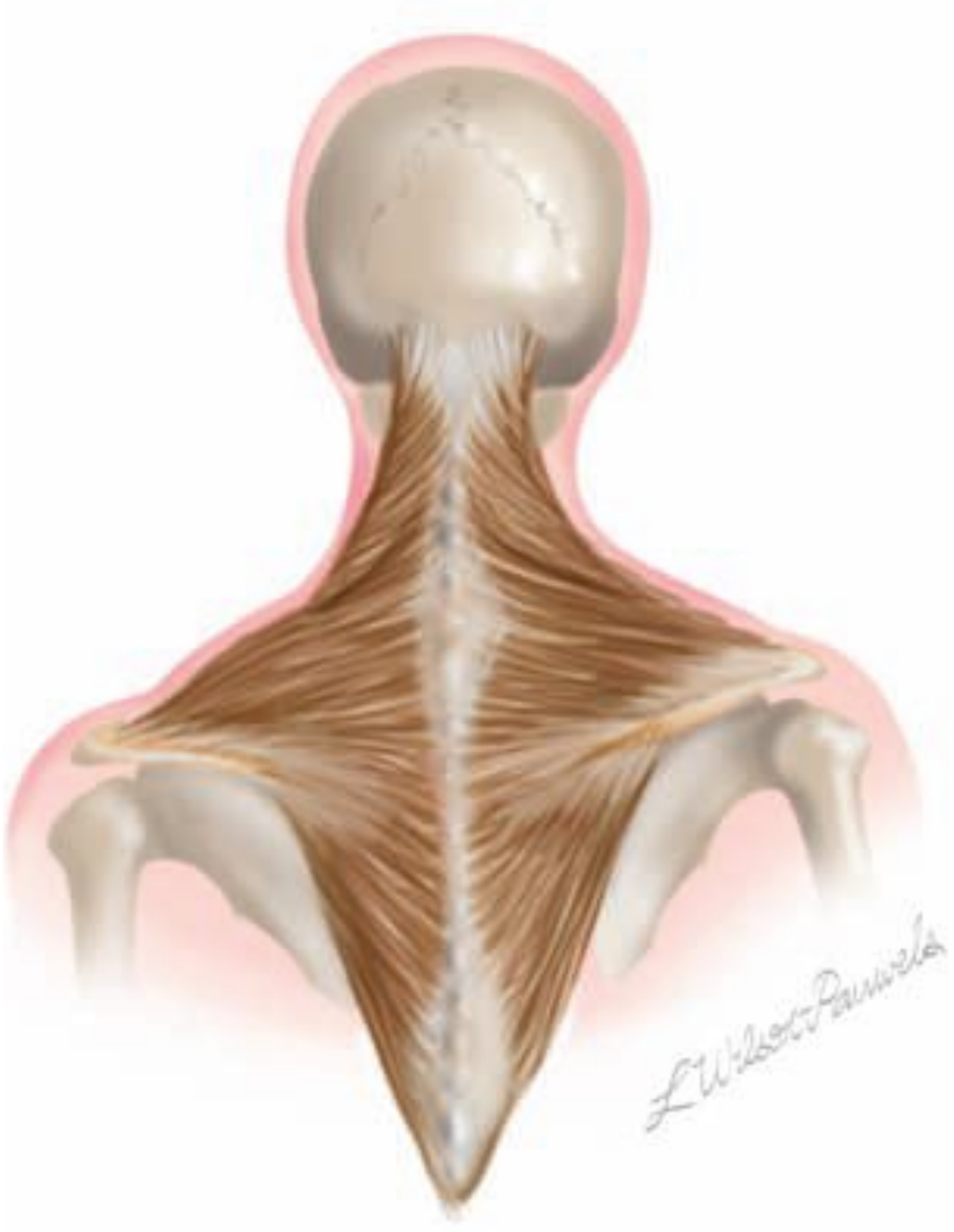

FIGURE XI–6 Left shoulder drop resulting from loss of action of upper fibers of the left trapezius muscle.

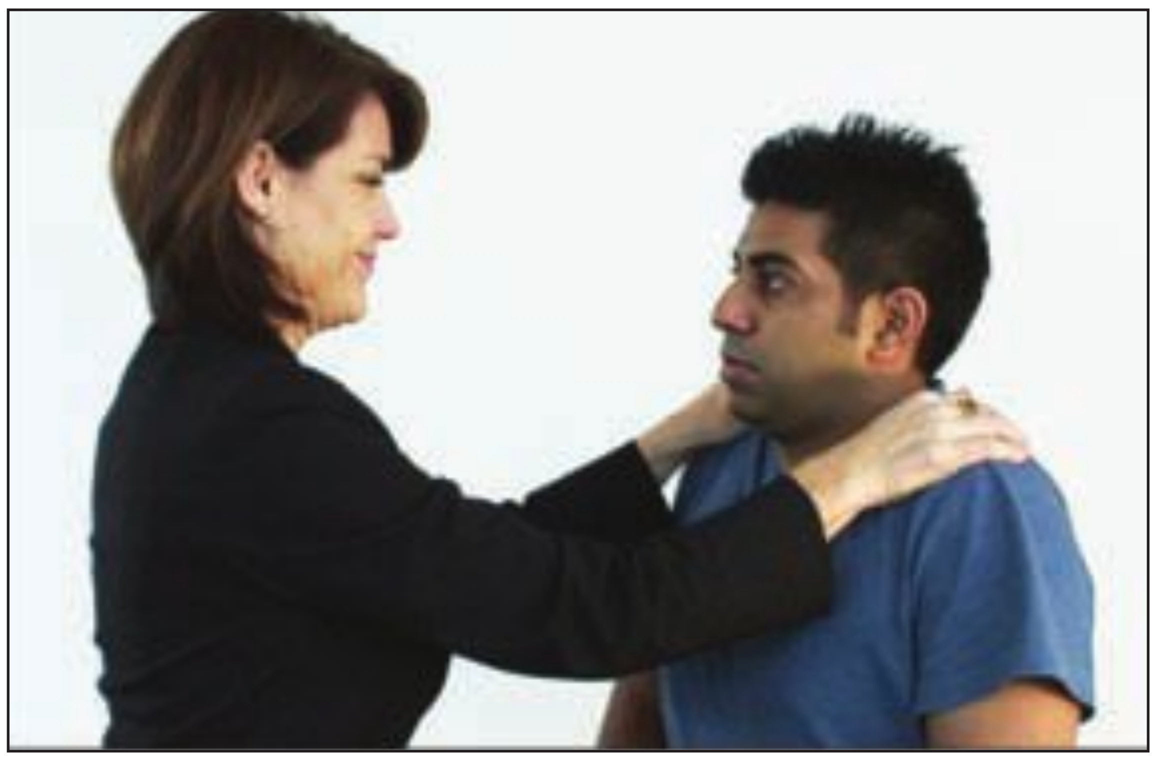

FIGURE XI–7 Patient shrugging his shoulders against resistance.

The upper fibers of the trapezius muscle are responsible for elevation and upward rotation of the scapula enabling abduction of the arm beyond 90 degrees. To assess the strength, ask the patient to abduct the arm through 180 degrees. The examiner applies resistance to the outstretched arm once the arm is beyond 90 degrees. The trapezius can also be tested by asking the patient to shrug the shoulders against resistance (Fig. XI–7). However, make sure the patient is not bracing the hands against the legs to keep the trapezius muscle elevated.

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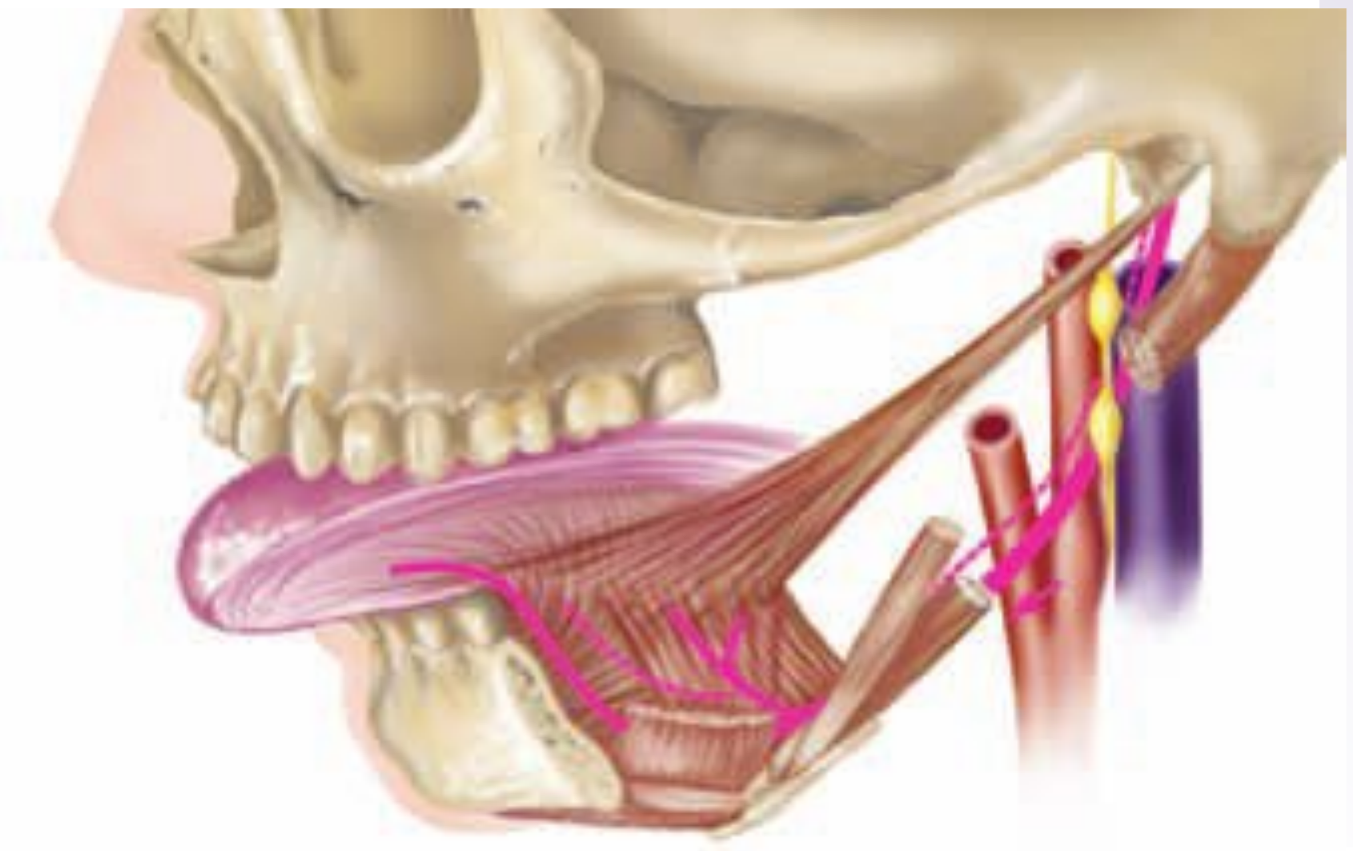

XII

Hypoglossal Nerve

CASE HISTORY

Todd is a 34-year-old bodybuilder. One afternoon while lifting weights at the gym, he experienced a sudden onset of pain in the left side of his neck radiating to his head. He had no other symptoms, but the pain was sufficient to make him stop his workout. The next day, the pain had almost disappeared, but over breakfast, he noticed he had difficulty moving food around in his mouth, his tongue felt heavy, and his speech was slurred.

Concerned that he had experienced a stroke, Todd went to the hospital. When examined, his eye movements and pupillary reflexes were normal. All his cranial nerves (CNs) functioned normally except CN XII. When he was asked to protrude his tongue, it deviated to the left. He was then asked to push his tongue into his right cheek and hold it there. The doctor was able to push the tongue toward the midline. Todd's taste and general sensation of the tongue were intact. All his other motor and sensory functions were normal.

A computed tomography (CT) scan of Todd's head was normal. An angiogram was done to look at blood flow to the brain. This test showed that Todd had suffered a dissecting aneurysm of the left internal carotid artery, which narrowed the lumen, thereby restricting blood flow and, in addition, caused a bulging of the vessel wall outward. However, the angiogram also demonstrated that the left cerebral hemisphere was still well perfused with blood from the collateral blood supply via the circle of Willis.

ANATOMY OF THE HYPOGLOSSAL NERVE

The rootlets of the hypoglossal nerve emerge from the anterior surface of the medulla in the ventrolateral sulcus between the pyramid and the olive. They converge to form two groups of rootlets that pierce the dura and then converge to form the hypoglossal nerve, which exits the cranium through the hypoglossal (anterior condylar) foramen in the posterior cranial fossa. After exiting the skull, the nerve courses medial to CNs IX, X, and XI. It passes laterally and downward close to the posterior surface of the inferior ganglion of the vagus nerve to lie between the internal carotid artery and the internal jugular vein and deep to the posterior belly of the digastric muscle. Crossing lateral to the bifurcation of the common carotid artery, the nerve loops anteriorly above the greater cornu of the hyoid bone. After passing deep to the intermediate tendon of the digastric, stylohyoid, and free posterior border of the mylohyoid muscles, the hypoglossal nerve runs on the lateral surface of the hyoglossus muscle. It then passes forward on the lateral surface of the genioglossus muscle, dividing to supply its target muscles in the tongue (Figs. XII–1 and XII–2 and Table XII–1).

The hypoglossal nerve supplies all but one of the extrinsic muscles of the tongue, the exception being the palatoglossus muscle, which is supplied by CN X, and all the intrinsic tongue muscles. The extrinsic muscles act to pull the tongue forward and protrude it (genioglossus), elevate and retract the tongue (styloglossus), depress the tongue (hyoglossus), as well as move it from side to side. The intrinsic muscles act to change the shape of the tongue.

The tongue has two very important functions. The phylogenetically “old” function is concerned with eating and initiating swallowing by the tongue pushing the bolus of food from the mouth into the oropharynx. These actions occur in response to sensory signals from the mouth carried in CN V. Gustatory (taste) and tactile (touch) signals carried in CNs V, VII, and IX pass from the mouth through the nucleus of the tractus solitarius, the trigeminal nucleus, and the reticular formation to act on the hypoglossal nucleus resulting in reflex activities such as swallowing, sucking, and chewing. The intricate and complex movements of the tongue in speech constitute the phylogenetically “new” function of the tongue.

Information from the inferior frontal cortex, premotor association cortex, and other cortical areas is projected to the primary motor area (precentral gyrus) of the cortex, which, in turn, sends signals to the hypoglossal nuclei via the corticobulbar tracts. Most of these projections are bilateral, with one exception: the cortical neurons that drive the genioglossus muscles project only to the contralateral hypoglossal nucleus.

The hypoglossal nucleus (Fig. XII–3) is composed of lower motor neurons whose axons form the hypoglossal nerve. It is located in the tegmentum of the medulla between the dorsal vagal nucleus and the midline (see Fig. XII–3A). It is a long, thin nucleus that is approximately coextensive with the olive (see Fig. XII–3B). The hypoglossal nucleus extends rostrally and forms a swelling in the distal part of the open medulla called the “hypoglossal triangle” (see Fig. XII–3C). Axons from the hypoglossal nucleus pass ventrally to the lateral side of the medial lemniscus to emerge as a number of rootlets in the ventrolateral sulcus between the olive and the pyramid (see Fig. XII–3D). Because the hypoglossal nuclei are located very close together, a nuclear lesion tends to affect both nuclei, causing bilateral loss of innervation to the tongue.

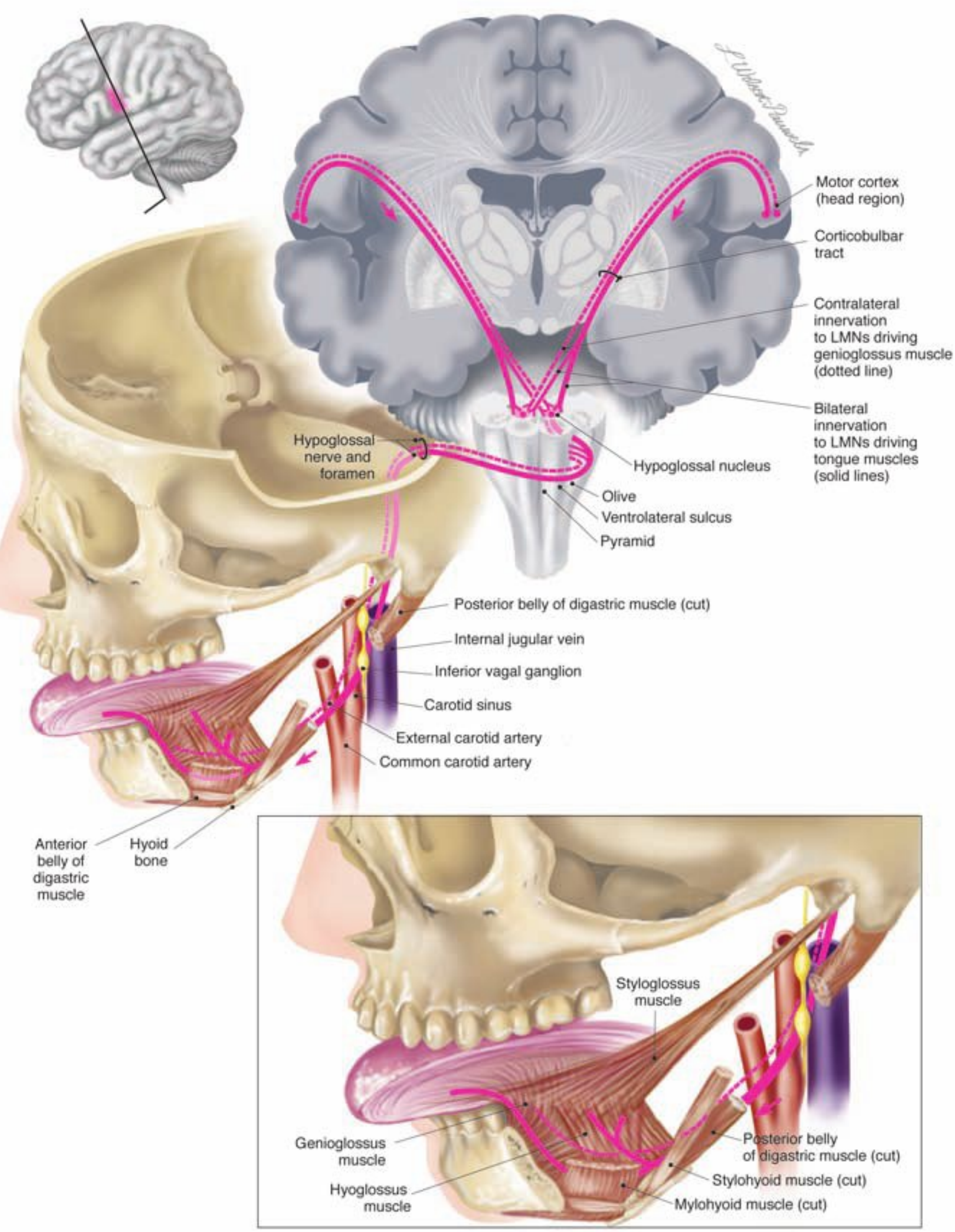
Motor cortex (head region)
Corticobulbar tract
Contralateral innervation to LMNs driving genioglossus muscle (dotted line)
Bilateral innervation to LMNs driving tongue muscles (solid lines)
Hypoglossal nerve and foramen
Hypoglossal nucleus
Olive
Ventrolateral sulcus
Pyramid
Posterior belly of digastric muscle (cut)
Internal jugular vein
Inferior vagal ganglion
Carotid sinus
External carotid artery
Common carotid artery
Anterior belly of digastric muscle
Hyoid bone
Styloglossus muscle
Genioglossus muscle
Hyoglossus muscle
Posterior belly of digastric muscle (cut)
Stylohyoid muscle (cut)
Mylohyoid muscle (cut)

FIGURE XII–1 Somatic motor component of the hypoglossal nerve (cranial nerve XII).

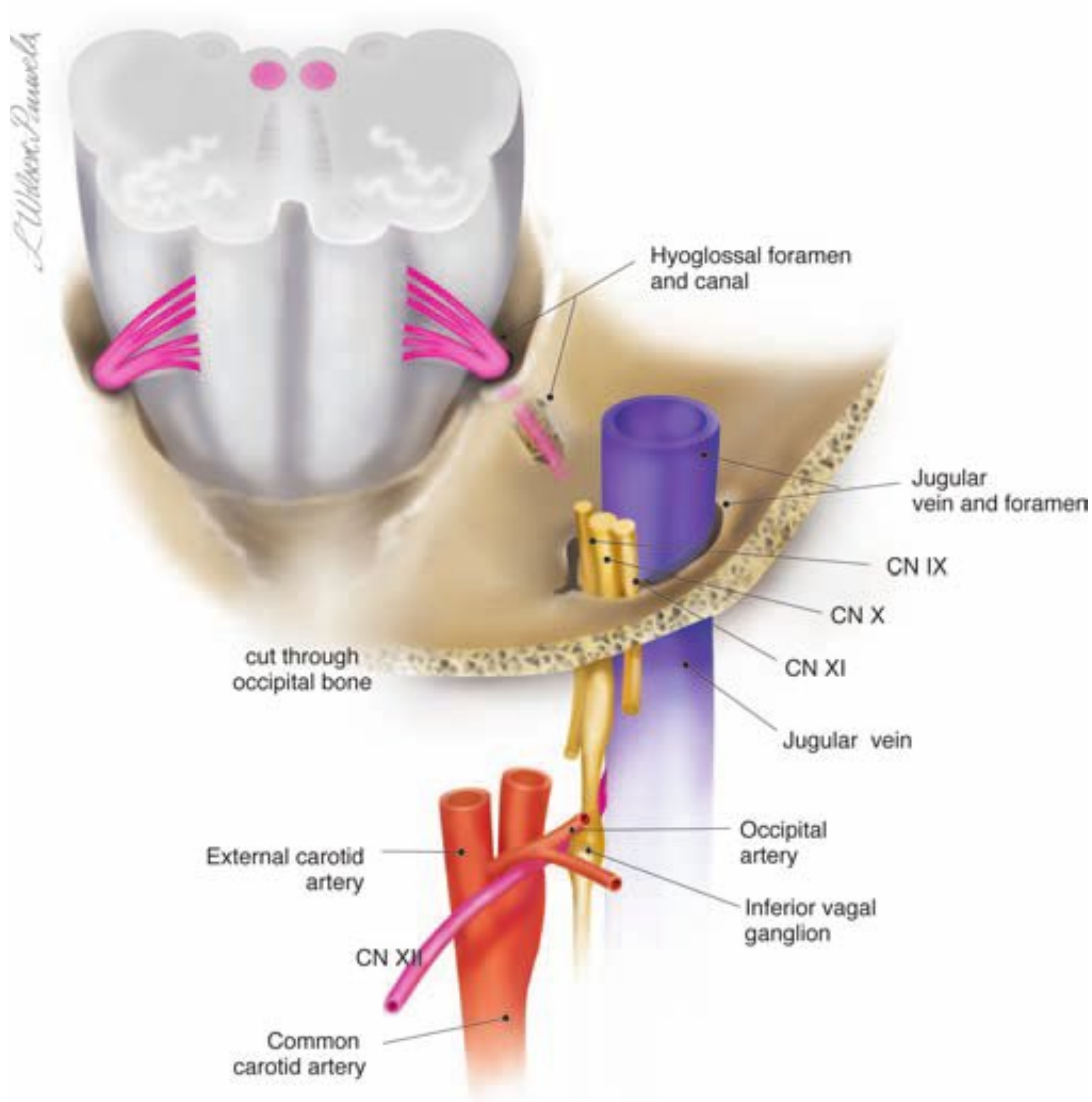

FIGURE XII–2 Route of the hypoglossal nerve through the hypoglossal canal to the bifurcation of the internal and external carotid artery

TABLE XII–1 Component, Nucleus and Function of the Hypoglossal Nerve (CN XII)

Component	Nucleus	Function
Somatic motor (efferent)	Hypoglossal nucleus	To innervate three of the four extrinsic muscles of the tongue (i.e., genioglossus, styloglossus, and hyoglossus) and all intrinsic muscles of the tongue

CASE HISTORY GUIDING QUESTIONS

1. How could Todd have damaged CN XII?
2. What CNs could be damaged by the process of internal carotid artery dissection?
3. Why could Todd still experience general sensation and taste from his tongue?
4. How would you differentiate between an upper motor neuron lesion (UMNL) and a lower motor neuron lesion (LMNL) of the hypoglossal nerve?

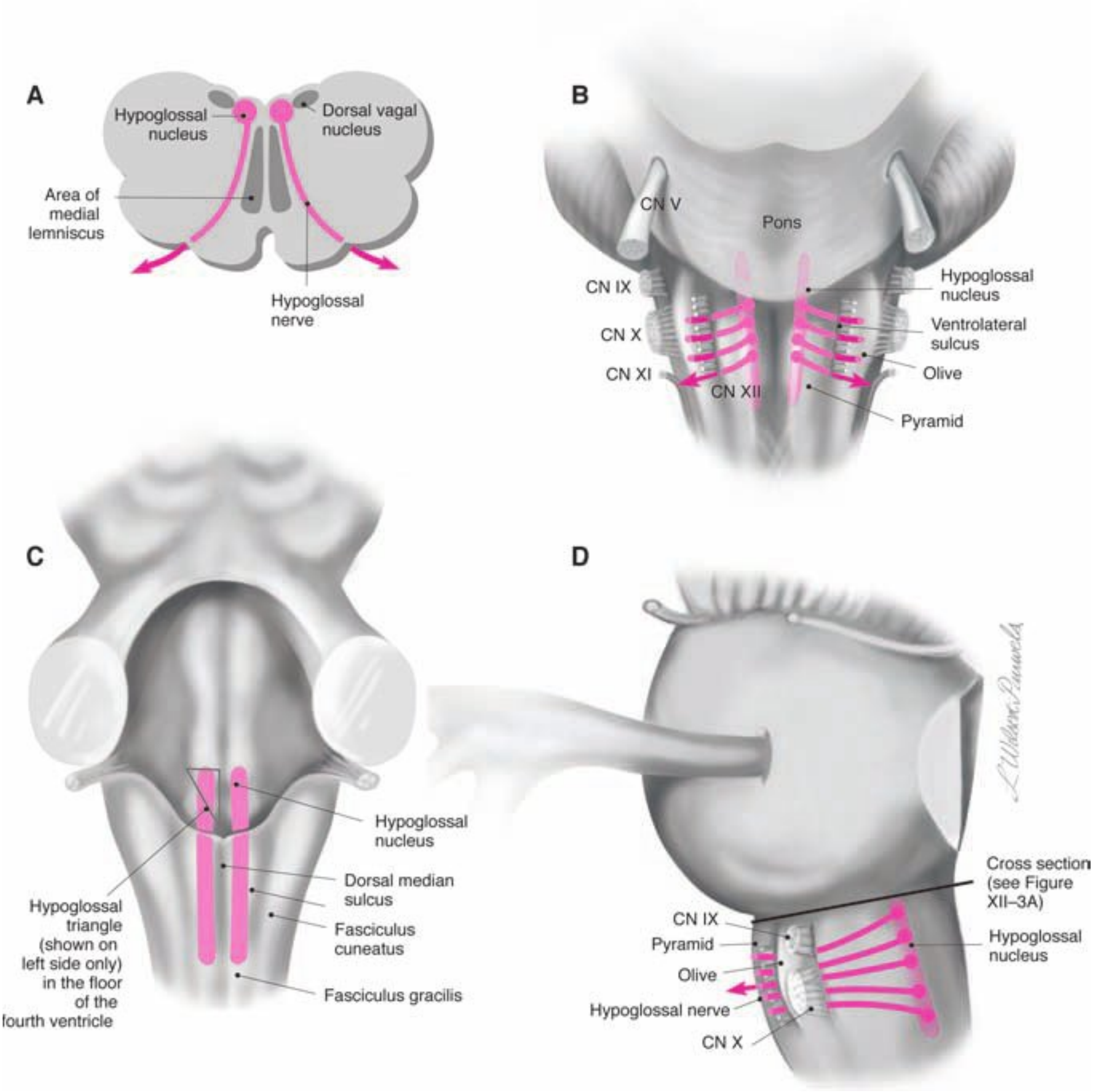

FIGURE XII–3 Views of the hypoglossal nucleus. **A**, Cross-section through the medulla–open portion. **B**, Ventral view. **C**, Dorsal view. **D**, Lateral view.

5. What findings confirmed that Todd had an LMNL?
6. Why were there no signs of fasciculations and atrophy of Todd's tongue?

1. How could Todd have damaged CN XII?

While straining lifting weights, Todd tore the inner lining (tunica intima) of his left internal carotid artery. Because of the tear, blood flowed between the tunica intima and the tunica media of the artery wall and formed a blood clot. This splitting of the layers of the vessel wall is referred to as an “arterial dissection” (Fig. XII–4). The blood clot narrowed the lumen of the vessel, decreasing blood flow, and significantly expanded the circumference of the internal carotid artery, thereby compressing the adjacent CN XII where it crossed the artery (see Fig. XII–4).

2. What CNs could be damaged by the process of internal carotid artery dissection?

Any nerves that lie in close proximity to the internal carotid artery could be affected. The most commonly affected are CNs IX, X, and XII. Isolated CN palsies such as Todd’s are a rare presentation and can be associated with an internal carotid artery dissection. More commonly, an internal carotid artery dissection results in occlusion of the artery

3. Why could Todd still experience general sensation and taste from his tongue?

Taste sensation from the tongue is carried primarily by CNs VII and IX, while general sensation from the tongue is carried in the mandibular division of CN V and

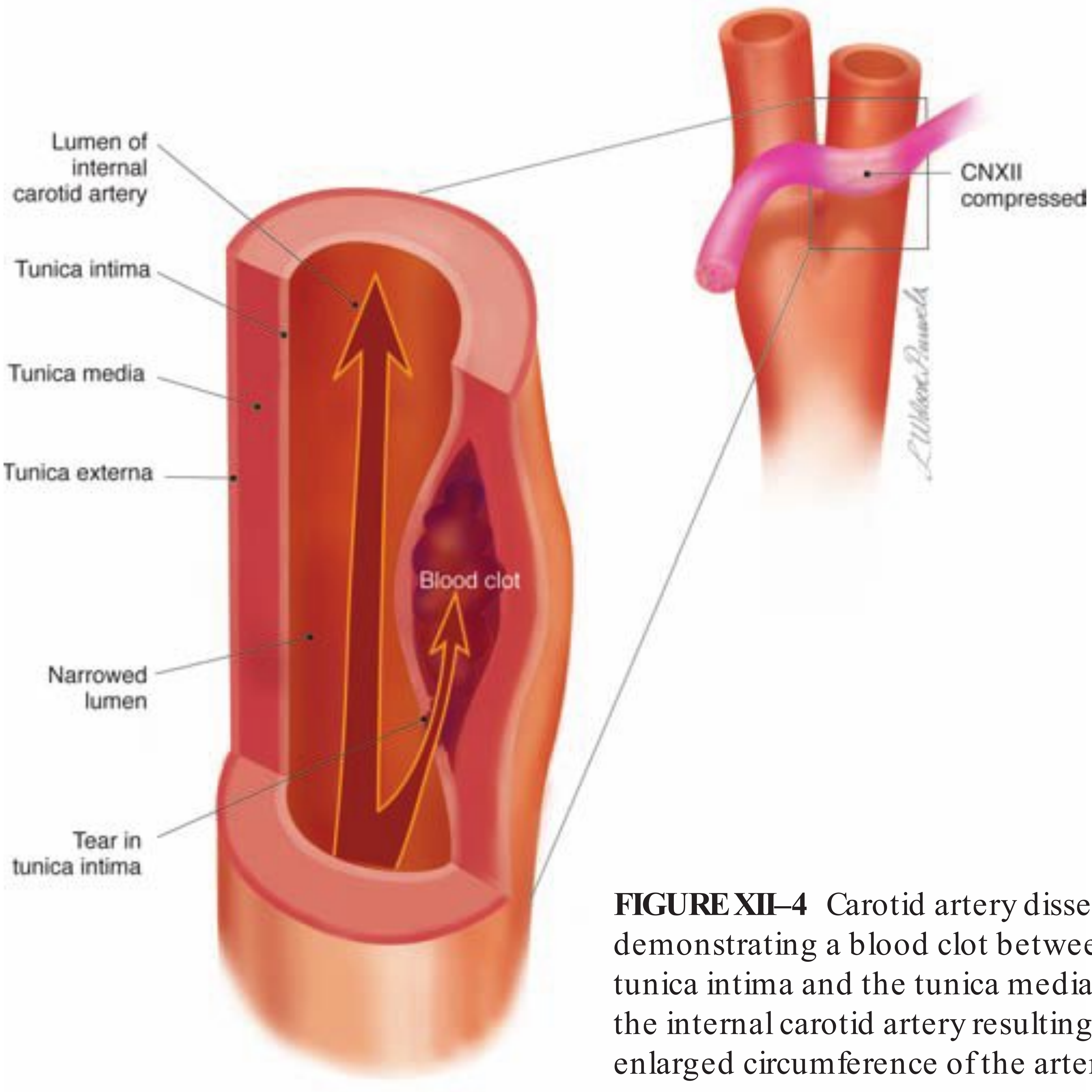

FIGURE XII–4 Carotid artery dissection demonstrating a blood clot between the tunica intima and the tunica media of the internal carotid artery resulting in an enlarged circumference of the artery.

CN IX. Both taste and general sensation were spared because the lesion is below the level of emergence of CNs V, VII, and IX.

4. How would you differentiate between a UMNL and an LMNL of the hypoglossal nerve?

The genioglossus is one of the most clinically important extrinsic tongue muscles because, unlike the other tongue muscles, it does not receive bilateral innervation. When the genioglossi muscles contract together, the tongue sticks straight out (Fig. XII–5). If one genioglossus muscle can contract and the other cannot, the tongue will deviate toward the inactive side.

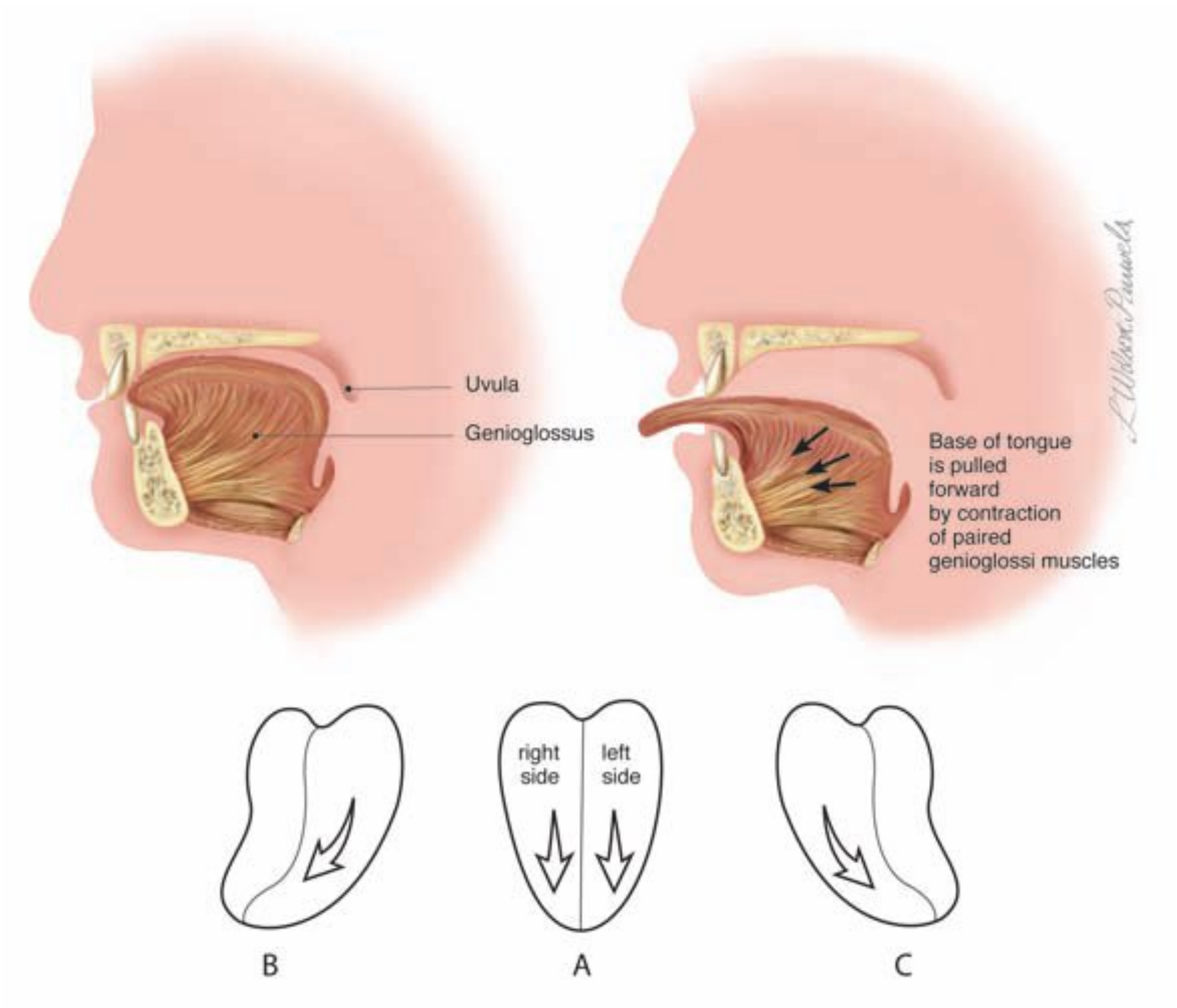
Uvula
Genioglossus
Base of tongue is pulled forward by contraction of paired genioglossi muscles
right side
left side
B
A
C

FIGURE XII–5 Action of the genioglossus muscle in sticking out the tongue.

- **A.** Balanced action of both genioglossus muscles is required to stick the tongue out in the midline
- **B.** When the right genioglossus muscle is weak or paralyzed, the left genioglossus muscle pushes the tongue to the weak side.
- **C.** When the left genioglossus muscle is weak or paralyzed, the right genioglossus muscle pushes the tongue to the weak side.

Damage to the upper motor neuron (UMNL) anywhere along the path of the axon from the cerebral cortex to the contralateral hypoglossal nucleus might result in paralysis of the contralateral genioglossus muscle. In this case, the tongue would deviate to the side opposite to the lesion owing to the unopposed action of the functioning genioglossus muscle (Fig. XII–6).

When the lower motor neuron is damaged (LMNL) anywhere between the hypoglossal nucleus and the tongue, ultimately there is flaccid paralysis of the ipsilateral half of the tongue with fasciculation and atrophy of tongue muscles on the affected side. In this case, the ipsilateral genioglossus muscle would be paralyzed and the tongue would deviate to the same side as the lesion (Fig. XII–7).

5. What findings confirmed that Todd had an LMNL?

Two findings confirmed that Todd had an LMNL: (1) The angiogram demonstrated that Todd had a dissection in the wall of the left internal carotid artery where it could compress the left hypoglossal nerve (see Fig. XII–4) and (2) Todd's tongue deviated to the left side owing to the unopposed action of the intact right genioglossus muscle (see Fig. XII–7).

6. Why were there no signs of fasciculations and atrophy of Todd's tongue?

When a muscle is denervated, it usually takes several weeks for fasciculations and atrophy to be noticeable. Todd was seen the day after his accident; therefore, fasciculations and atrophy of his left tongue muscles would not have had time to develop.

CLINICAL TESTING

At Rest

Initially, the tongue is assessed by examining it at rest (Fig. XII–8A). Ask the patient to open the mouth and look for evidence of fasciculations or atrophy. At rest, if there is pathology present, the tongue may deviate to the unaffected side owing to the unopposed action of the styloglossus muscle that draws the tongue up and back (see Fig. XII–1). See also "Cranial Nerves" website.

During Protrusion

Next, ask the patient to protrude the tongue (see Fig. XII–8B). An LMNL will result in the tongue deviating to the ipsilateral side. The tongue deviates to the weak side because of the unopposed action of the intact contralateral genioglossus muscle. If the lesion involves upper motor neurons, the tongue will deviate to the contralateral side.

Lastly, ask the patient to push the tongue against the inside of the cheek (see Fig. XII–8C). If both genioglossi muscles are intact, the examiner, offering resistance by pushing on the skin of the cheek, should not be able to move the tongue to the midline from either cheek. A weakness in one of the genioglossus muscles will allow the examiner to push the tongue toward the weak side. See also "Cranial Nerves" website.

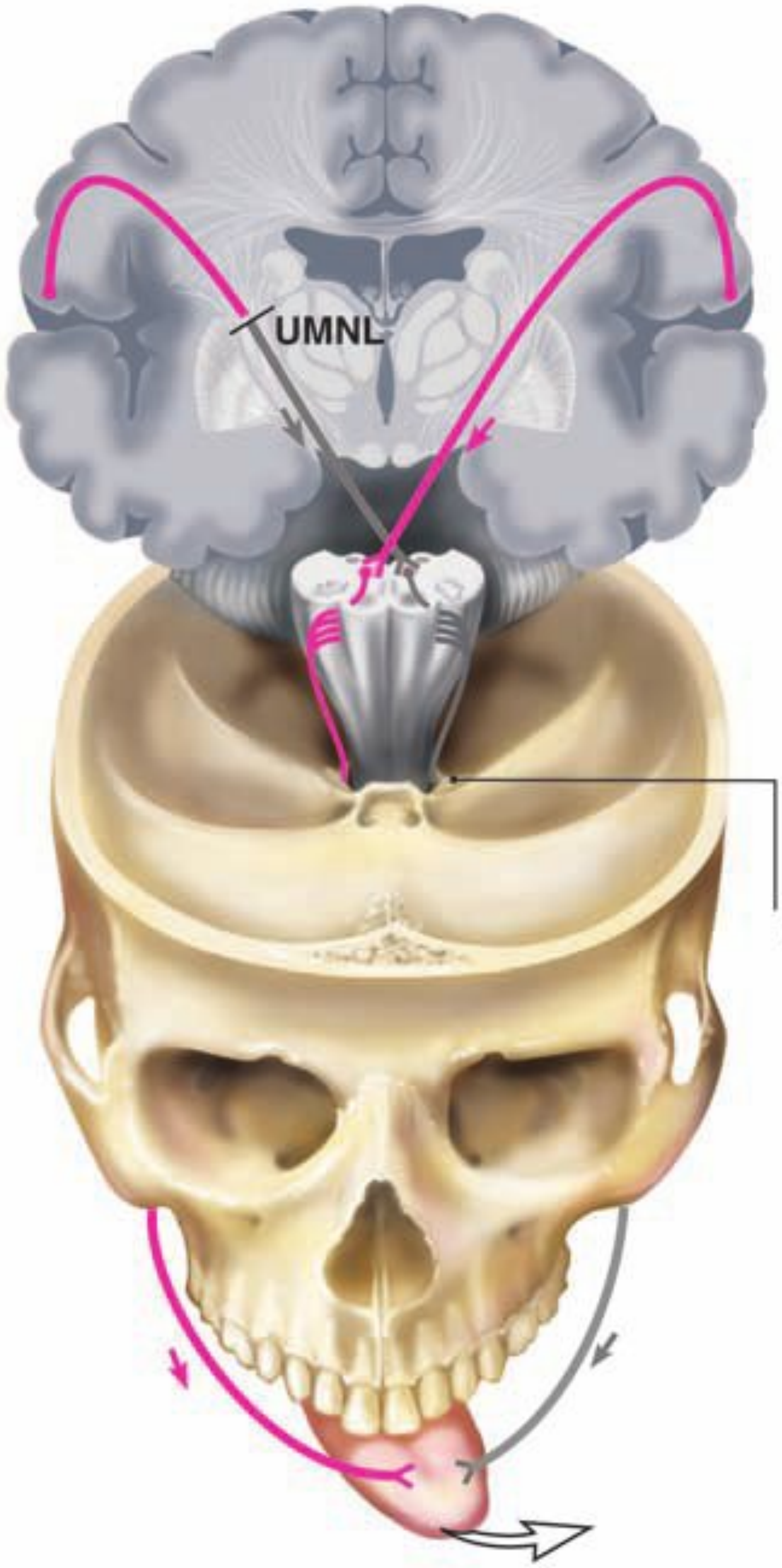

FIGURE XII–6 CNXII Upper Motor Neuron Lesion (UMNL).
Illustration demonstrates contralateral fibers to genioglossus muscles only.
In UMNLs. the protruded tongue deviates to the side contralateral to the lesion.

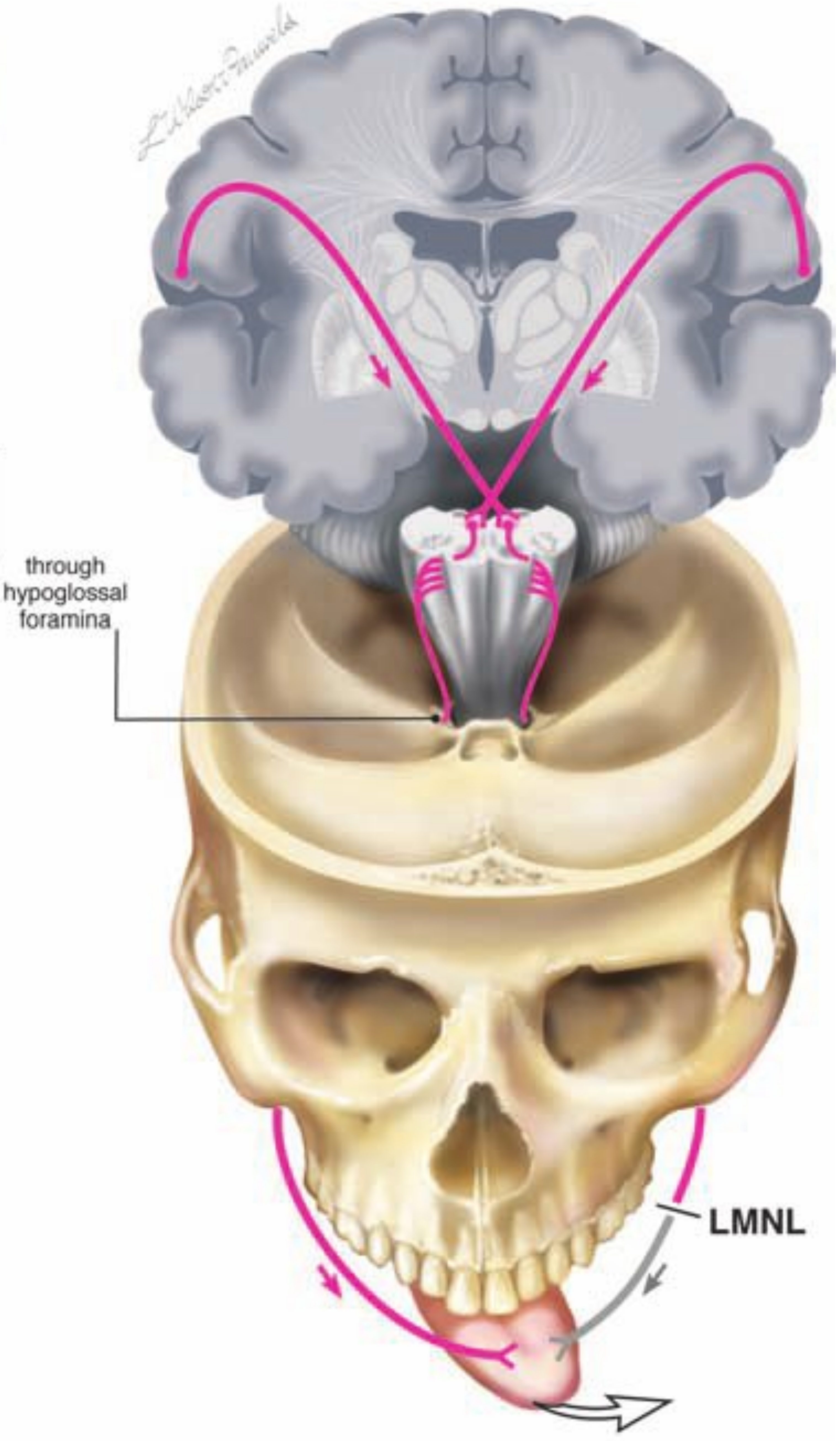

FIGURE XII–7 CNXII Lower Motor Neuron Lesion (LMNL).
Illustration demonstrates contralateral fibers to genioglossus muscles only.
In LMNLs, the protruded tongue deviates to the same side as the lesion (this is Todd's lesion).

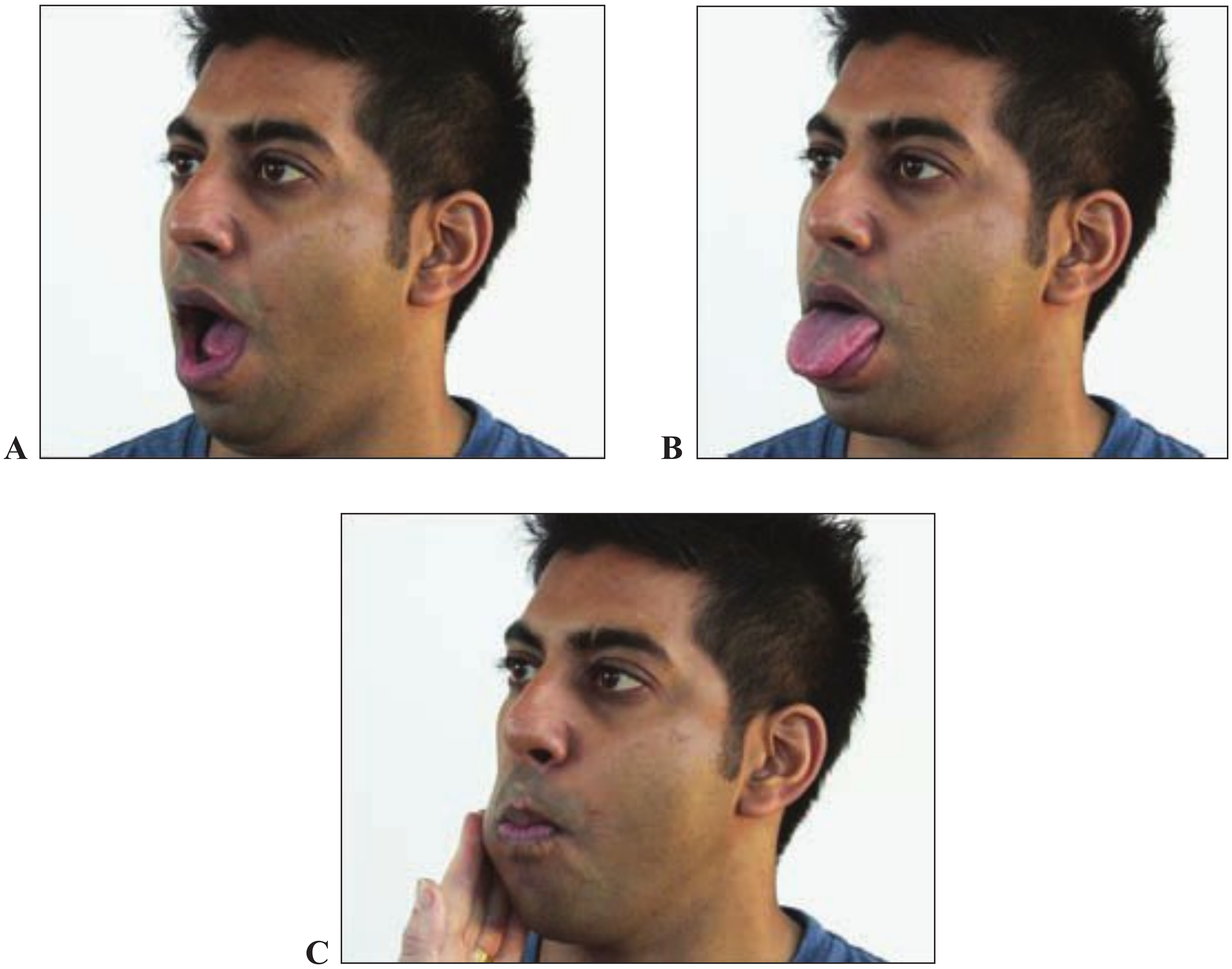

FIGURE XII–8 Testing of the tongue involves; **A**, tongue at rest, frontal view; **B**, protruding the tongue to observe if it sticks straight out or deviates to the side; and **C**, pushing the tongue into the cheek against resistance.

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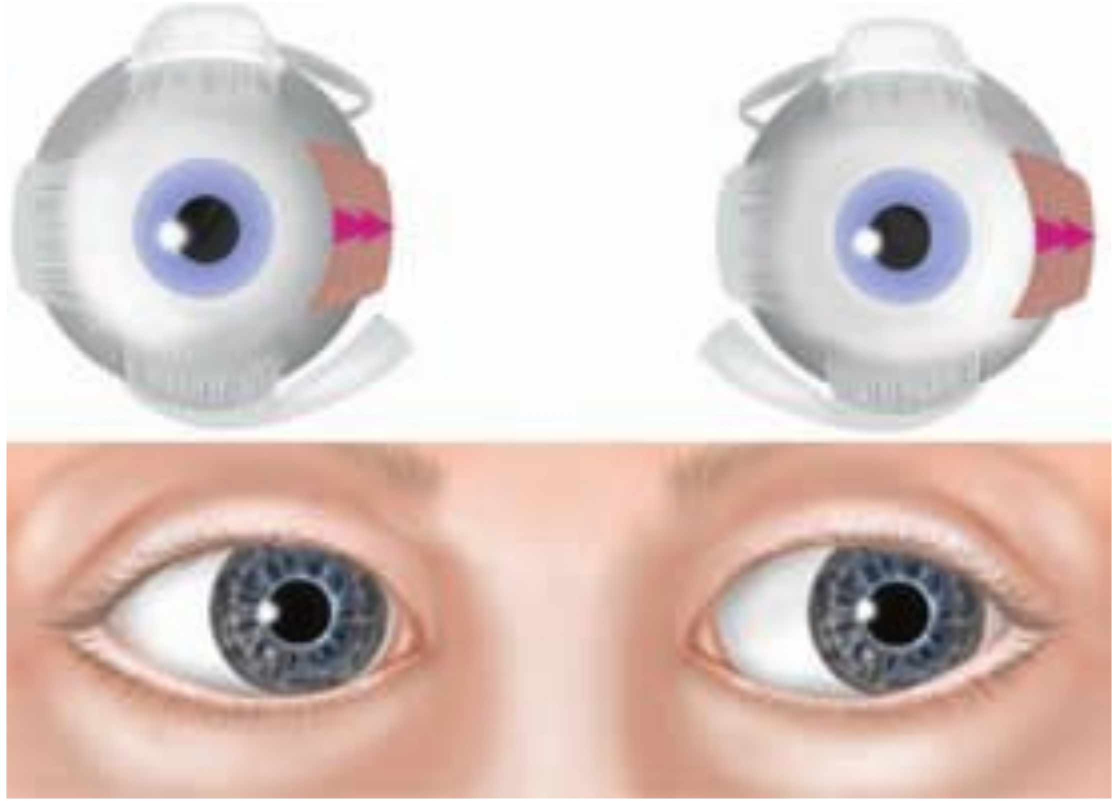


13

Coordinated Eye Movements and Clinical Testing of the Somatic Motor Components of Cranial Nerves III, IV, and VI

EYE MOVEMENTS

The human eye includes a specialized area called the “fovea,” which provides for high-resolution imaging and color discrimination (see Chapter II). The human oculomotor system acts to direct this specialized region toward objects of interest in the visual field and to maintain this direction. When the eye moves, the eye as a whole is not displaced. Rather, it rotates around three orthogonal axes that pass through the center of the globe (Fig. 13–1). For convenience in description, eye movements are described using the cornea as a reference point. “Abduction,” therefore, indicates that the cornea moves away from the nose, and “adduction” indicates that the cornea moves toward the nose. The corneas can be directed up or down in upward gaze and downward gaze. When the head is tilted laterally, the eyes rotate (incyclotorsion or excyclotorsion) in the opposite direction, compensating for a maximum of 40 degrees of head tilt.

The eye is moved by a total of six muscles whose actions are listed in Table 13–1. The amount that the eye can move in any one direction is limited by the attached extraocular muscles and optic nerve. Combining these movements allows the cornea to move in any direction to a maximum of 45 degrees from the

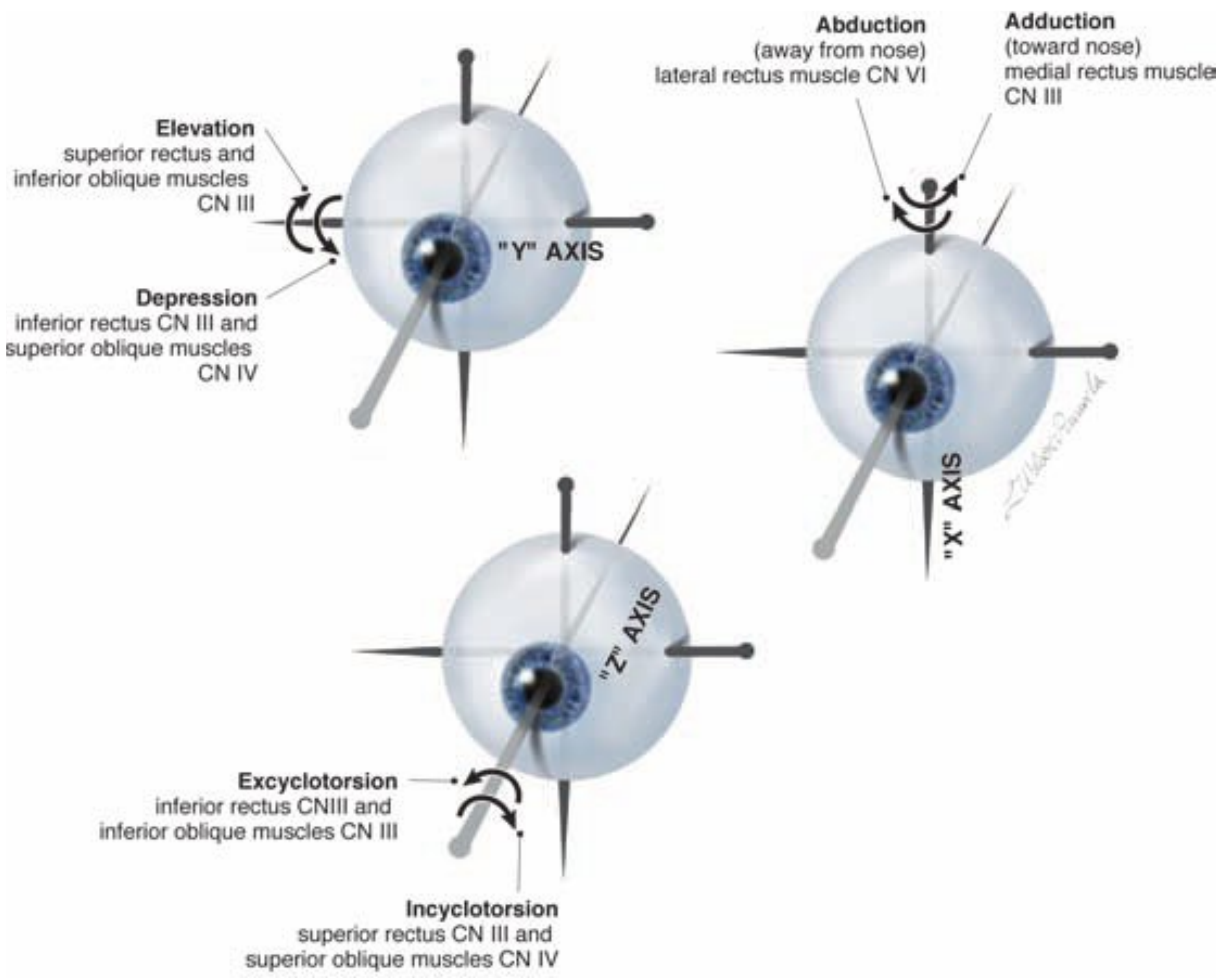


FIGURE 13–1 Right eye movements around the "X," "Y," and "Z" axes.

relaxed position. As a rule, however, the eye does not usually move to the extremes of its range, but remains within 20 degrees of the relaxed position. Figure 13–2 depicts the visual axis, which is an imaginary line connecting the object of fixation to the fovea. When the eye is at rest, the visual axis is parallel to the medial wall of the bony orbit.

TABLE 13–1 Actions of the Extraocular Muscles

Extraocular Muscle	CN	Primary Action	Secondary Action	Tertiary Action	Testing Position
Medial rectus	III	Adduction	None	None	Adduction
Lateral rectus	VI	Abduction	None	None	Abduction
Superior rectus	III	Elevation	Incyclotorsion	Adduction	Up and out
Inferior rectus	III	Depression	Excyclotorsion	Adduction	Down and out
Superior oblique	IV	Incyclotorsion	Depression	Abduction	Down and in
Inferior oblique	III	Excyclotorsion	Elevation	Abduction	Up and in

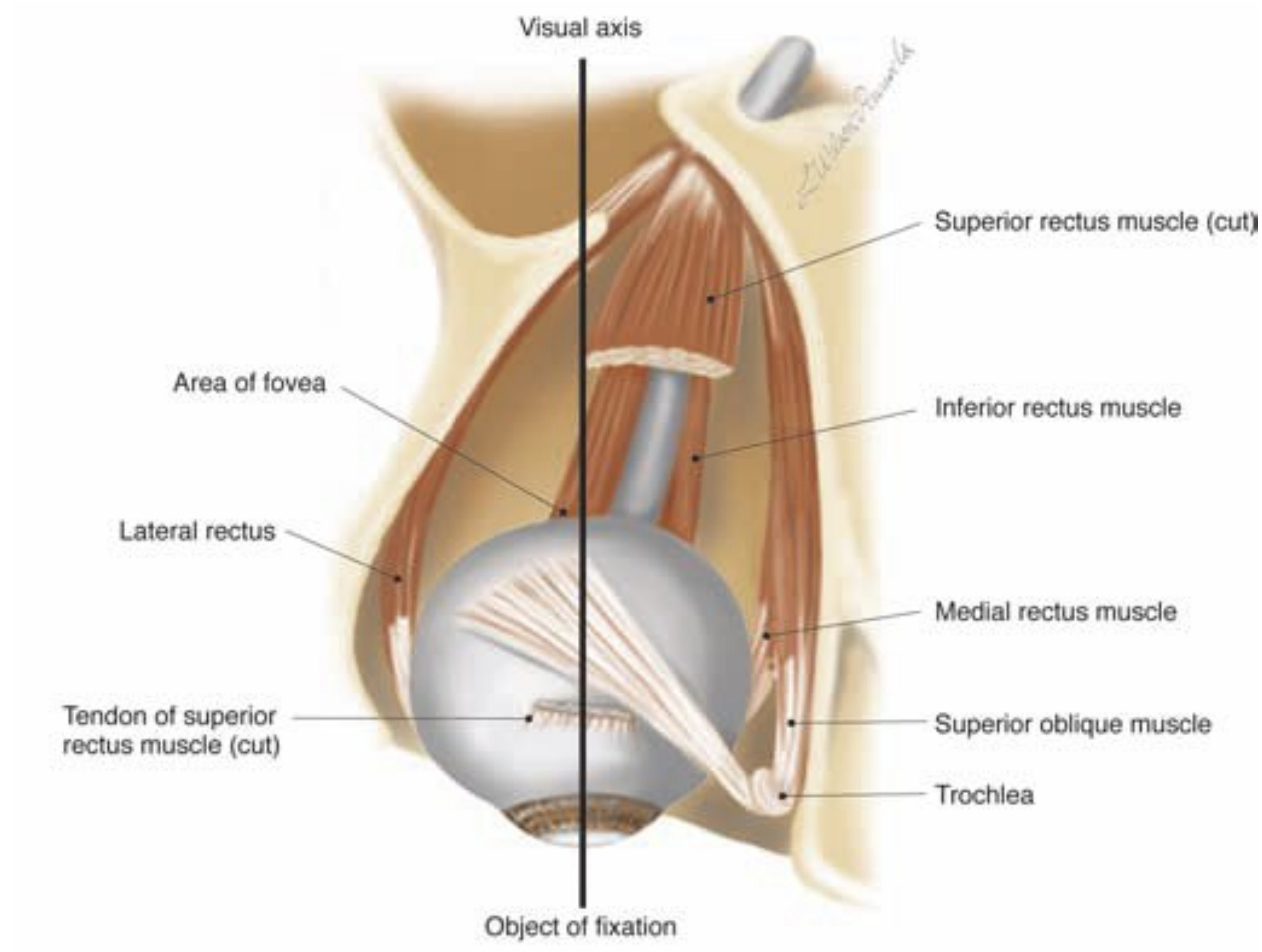


FIGURE 13–2 Superior view of the eye and orbit at rest. Superior rectus muscle has been cut to reveal the tendon of the superior oblique muscle. The visual axis, a line connecting the object of fixation and the fovea is indicated.

Eye movements are produced by the coordinated contraction and relaxation of the six extraocular muscles in each orbit. The action of the muscles is determined by the starting position of the eye in the orbit as well as by the origin and insertion of each muscle. The rectus muscles pass through connective tissue sheaths, also referred to as “pulleys,” located several millimeters posterior to the equator of the globe and approximately 1 cm posterior to the insertion of the rectus tendons. These pulleys limit side-slip movement of the rectus muscles during eye movement and act as their functional origin. Figure 13–3 illustrates this point.

Medial and Lateral Rectus Muscles

The medial and lateral rectus muscles are involved in horizontal movement. In conjugate horizontal gaze (wherein both eyes move in the same direction by the same amount), the medial rectus muscle (cranial nerve [CN] III) of one eye contracts in conjunction with the lateral rectus muscle (CN VI) of the other eye so that both eyes move either to the left or to the right. The medial rectus muscle adducts and the lateral rectus muscle abducts (Fig. 13–4). These actions are maintained even when the eye is elevated or depressed because of the constraining action of the connective tissue muscle sheaths (see Fig. 13–3).

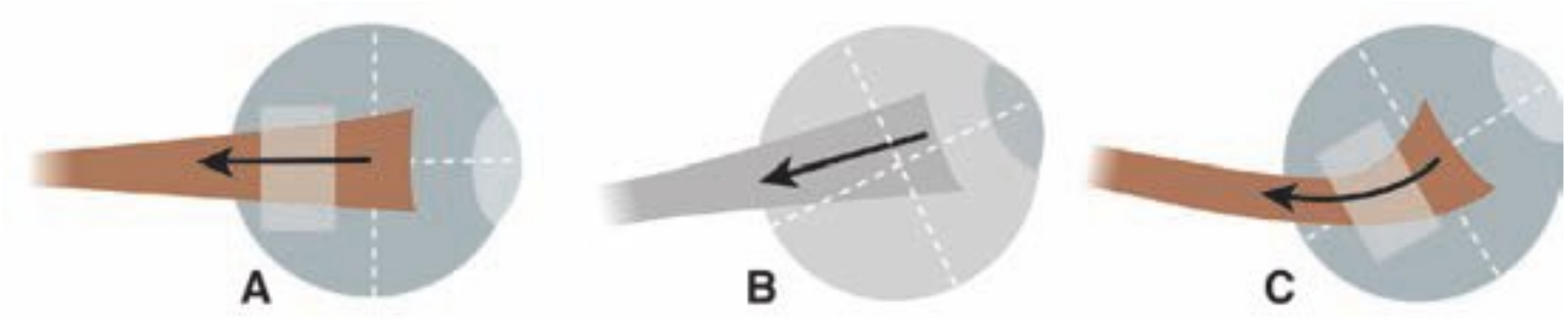


FIGURE 13–3 The action of the connective tissue muscle sheaths, also referred to as pulleys, in maintaining the direction of pull of the muscle. The lateral rectus muscle is depicted.

- **A.** In the primary position the pull of the lateral rectus muscle is along the horizontal meridian of the globe and therefore only abducts the eye.
- **B.** If the pulley were not present, when the eye is elevated (shown) or depressed, the muscle belly would tend to slide across the surface of the globe such that its pull would now be at an angle to the horizontal meridian, and it would contribute to elevation (shown) or depression, as well as abduction.
- **C.** The pulley restricts the movement of the muscle and ensures that its pull remains along the horizontal meridian of the eye.

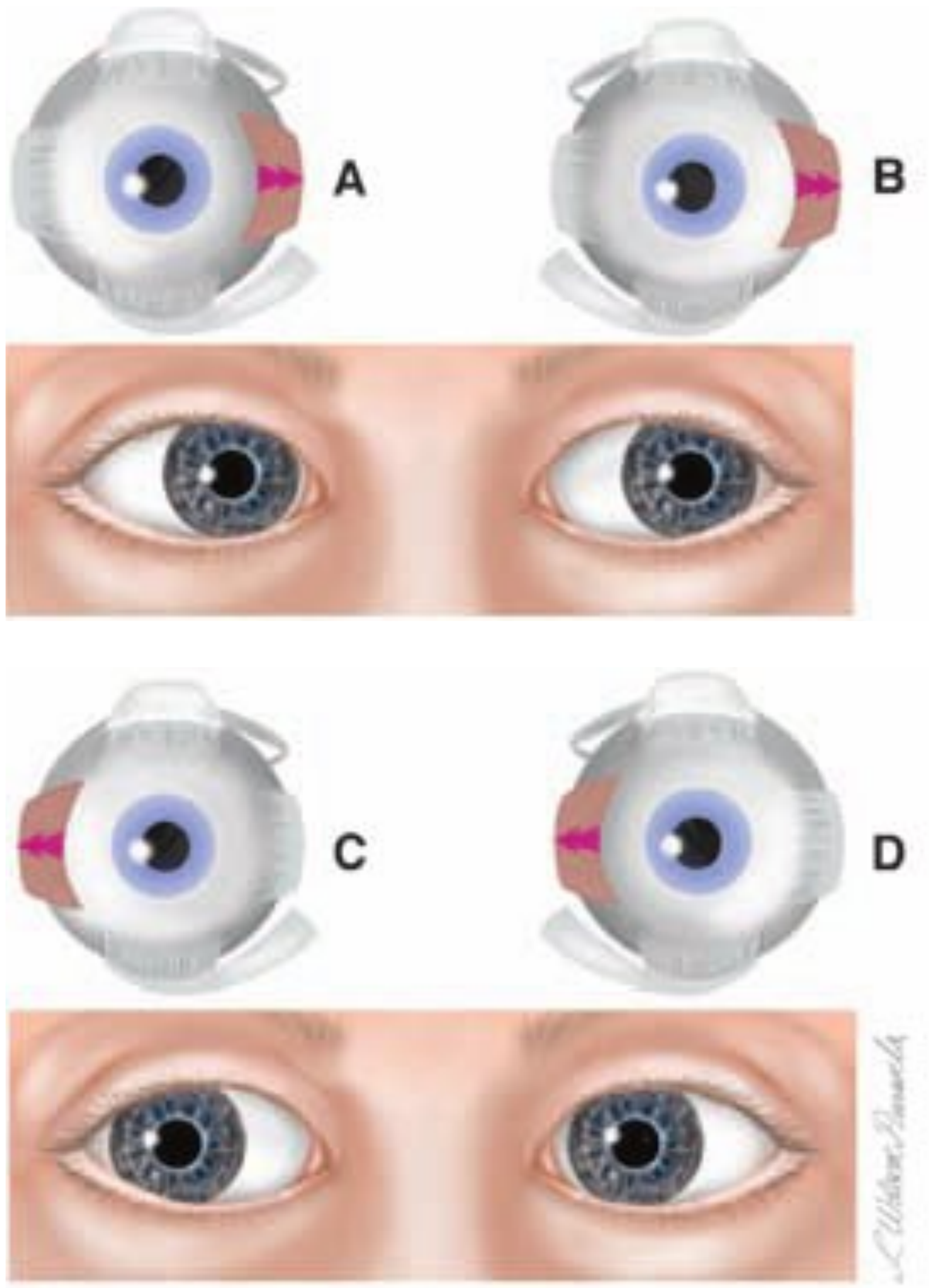


FIGURE 13–4 Left gaze requires the combined action of the A, right medial rectus muscle (CN III) and the B, left lateral rectus muscle (CN VI).

Right, gaze requires the combined action of the A, right lateral rectus muscle (CN VI) and the B, left medial rectus muscle (CN III).

Superior and Inferior Rectus Muscles

When the eye is abducted 23 degrees from the primary position, the superior and inferior rectus muscles are aligned with the visual axis and, therefore,elevate and depress the eye, respectively (Fig. 13–5A). In other eye positions, the pull of these muscles is at an angle to the visual axis, and as a result, they also contribute to both adduction and torsional movements (see Fig. 13–5B). The connective tissue sheaths

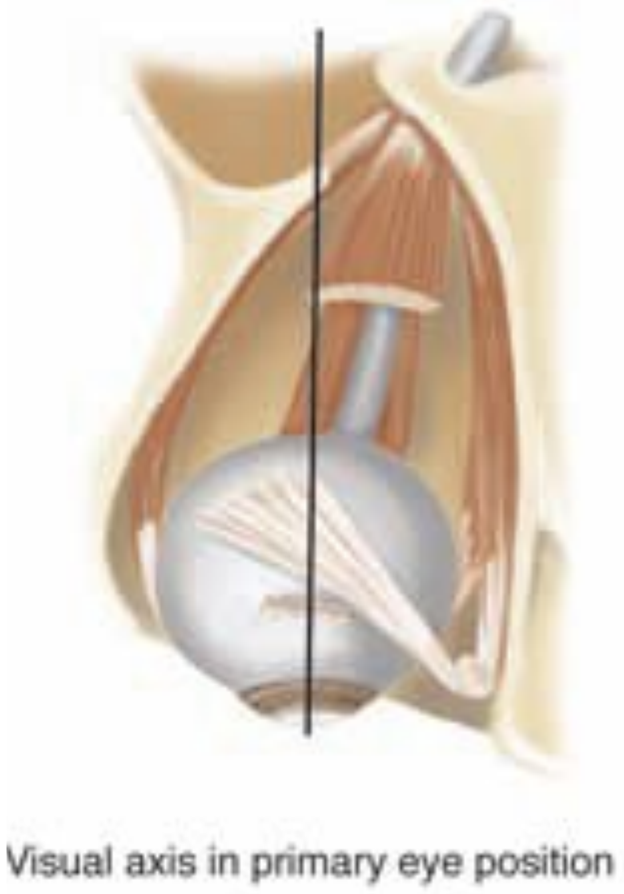
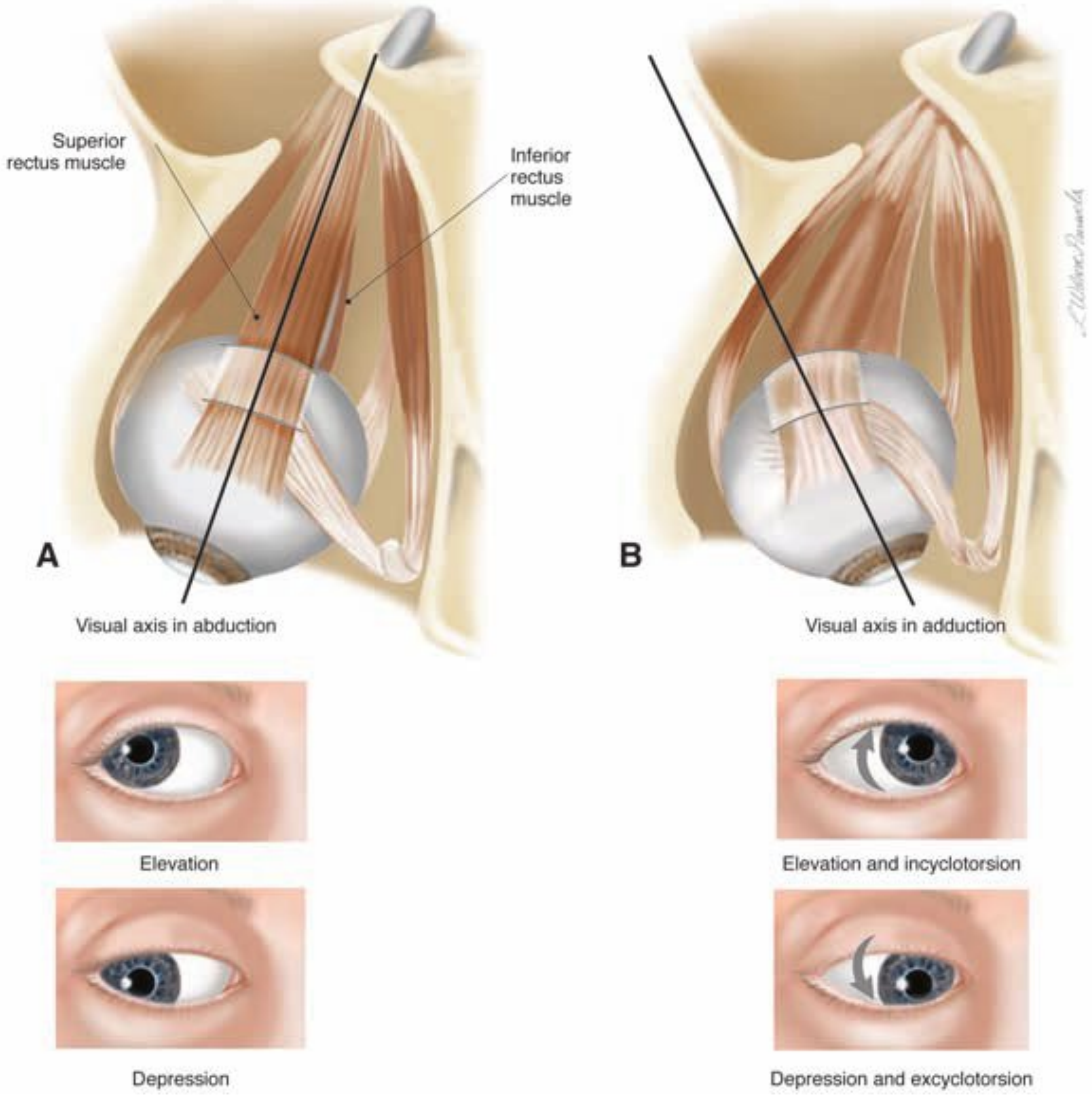


FIGURE 13–5

A. Superior and inferior rectus muscles in abduction. When the right eye is abducted, the pull exerted by the superior and inferior recti muscles (CN III) is parallel to the visual axis. Therefore, the superior and inferior rectus muscles **elevate** and **depress** the eye.

B. Superior and inferior rectus muscles in adduction. When the right eye is adducted, the pull of the superior and inferior rectus muscles is at an angle to the visual axis such that the eye is rotated around the visual axis (**incyclotorsion** and **excyclotorsion**). The connective tissue pulleys constrain this action somewhat.



that the rectus muscles pass through constrain their direction of pull, however, minimizing these secondary actions.

Superior and Inferior Oblique Muscles

Unlike the rectus muscles, the oblique muscles insert on the back of the globe. Their direction of pull lies at an angle of 51 degrees to 54 degrees to the visual axis. Their primary actions, therefore, are to cause rotation around the visual axis. The superior oblique muscle causes incyclotorsion and the inferior oblique muscle causes excyclotorsion. When the eye is abducted (away from the nose), they rotate the eye about the visual axis (Fig. 13–6). When the eye is fully adducted (toward the nose), the superior and inferior oblique muscles depress and elevate the eye, respectively (see Fig. 13–6). At eye positions between fully abducted and fully adducted, the superior and inferior oblique muscles contribute to both vertical and rotatory movement.

To test eye movements, draw a large "H" in the air a few feet in front of the patient and ask her or him to follow your finger with her or his eyes. The horizontal bar of the "H" will test medial and lateral rectus muscles. The two vertical bars of the "H" will isolate and test the motion of the superior or inferior rectus muscles and the inferior or superior oblique muscles. The eyes should move in a smooth, coordinated motion throughout the "H" (Fig. 13–7).

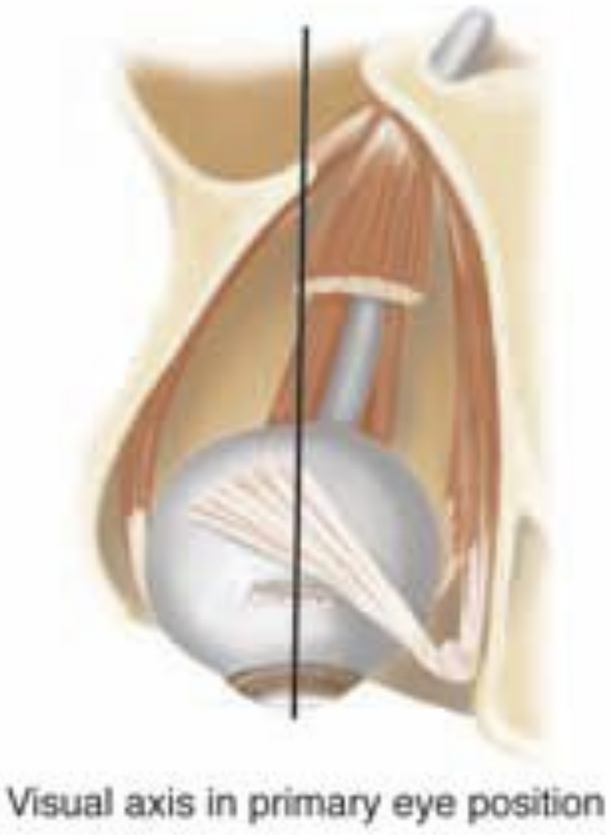
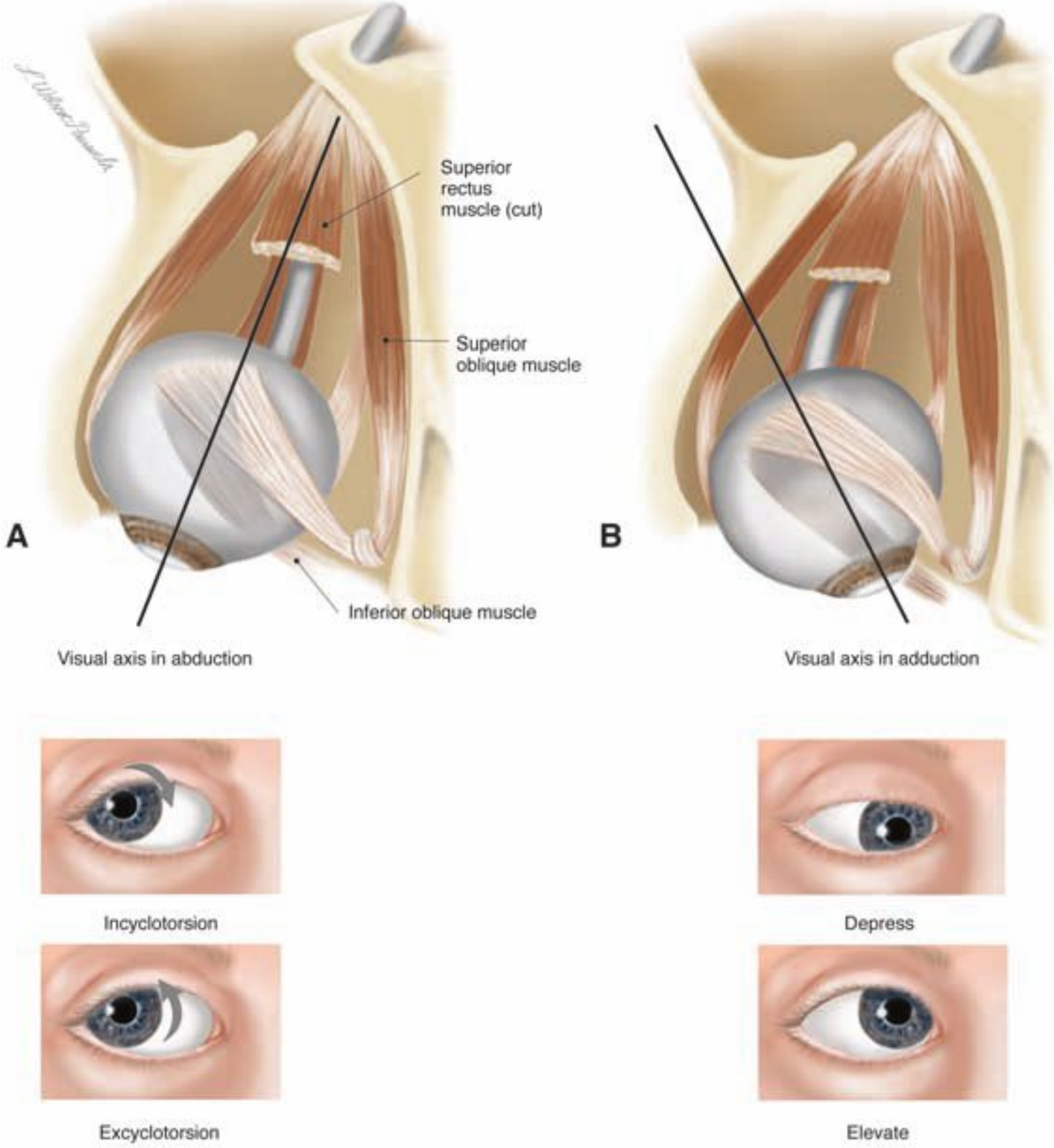


FIGURE 13–6

A. Superior and inferior oblique muscles in abduction. When the right eye is abducted, the pull exerted by the superior (CN IV) and inferior oblique (CN III) muscles is at an angle to the visual axis. Therefore, the superior and inferior oblique muscles **incyclotort** and **excyclotort** the eye, respectively.

B. Superior and inferior oblique muscles in adduction. When the right eye is adducted, the pull exerted by the superior and inferior oblique muscles is parallel to the visual axis. Therefore, the superior (CN IV) and inferior (CN III) oblique muscles **depress** and **elevate** the eye, respectively.



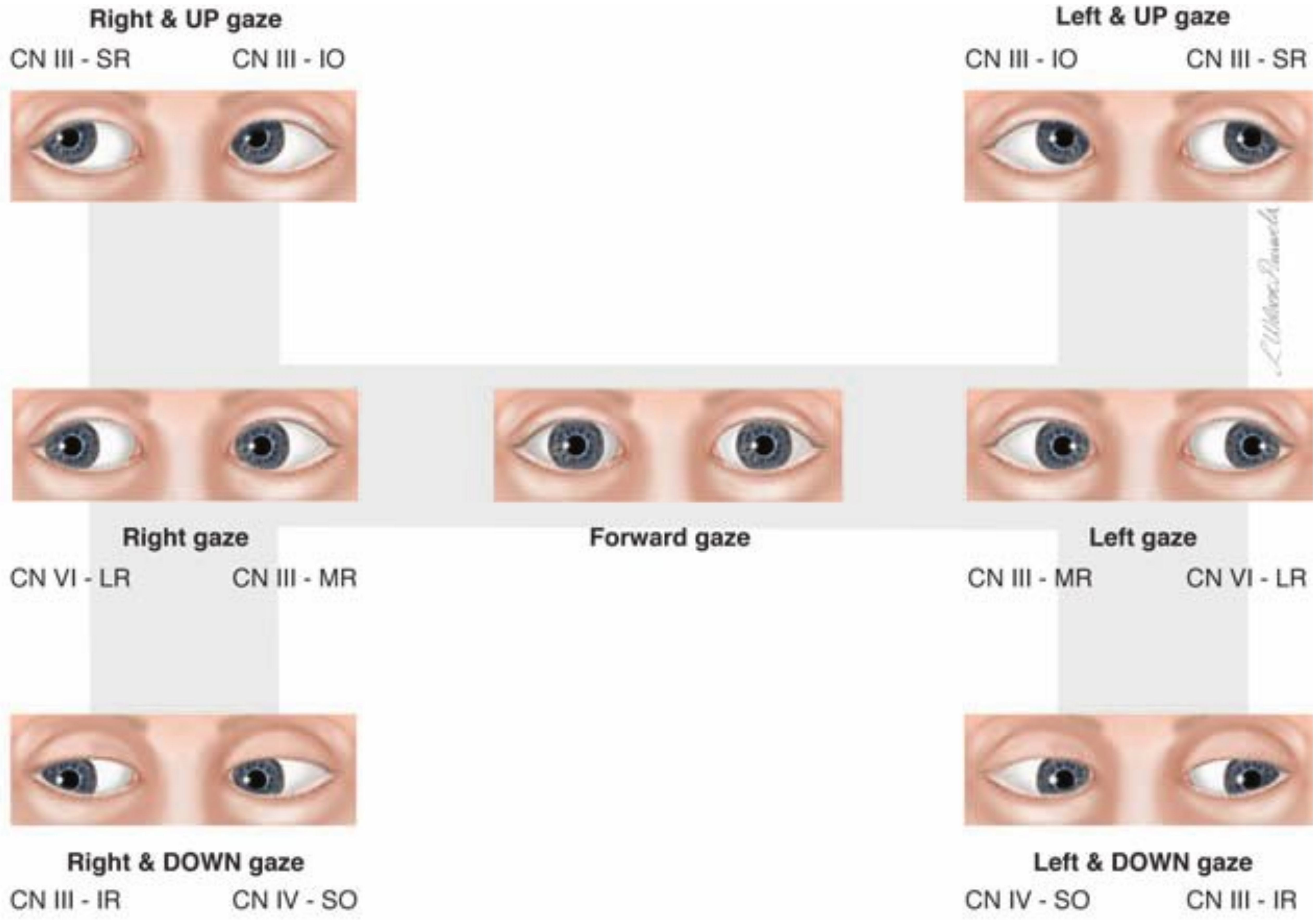


FIGURE 13–7 In conjugate eye movement, the "H" motion test provides a vehicle to assess the integrity of individual eye muscles. The patient is asked to follow the physician's finger as it moves horizontally and then up and down at each end of the "H". The physician performs the movement while observing one eye, and then repeats it while observing the other eye. The muscle in each eye whose action is being tested and the nerve that drives it are indicated at each position.
Muscle names: Superior Rectus (SR), Inferior Rectus (IR), Medial Rectus (MR), Lateral Rectus (LR), Superior Oblique (SO), Inferior Oblique (IO).

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